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Original Article

Process Improvement: A Multi-Registry Database Abstraction Success Story

Victor Abrich, MD; Roxann Rokey, MD, FACC, FASE; Christopher Devadas, BSE; Julie Uebel, RN, BSN

Abstract: Background: The St. Joseph Hospital/Marshfield Clinic Cardiac Database Registry submits data to the National Cardiovascular Data Registry (NCDR) and to the Society of Thoracic Surgeons (STS) National Database. Delayed chart abstraction is problematic, since hospital policy prohibits patient care clarifications made to the medical record more than 1 month after hospital discharge. This can also lead to late identification of missed care opportunities and untimely notification to providers. Our institution was 3.5 months behind in retrospective postdischarge case abstraction. A process improvement plan was implemented to shorten this delay to 1 month postdischarge. Methods: Daily demand of incoming cases and abstraction capacity were determined for 4 employees. Demand was matched to capacity, with the remaining time allocated to reducing backlog. Results: Daily demand of new cases was 17.1 hours. Daily abstraction capacity was 24 hours, assuming 6 hours of effective daily abstraction time per employee, leaving 7 hours per day for backlogged case abstraction. The predicted time to reach abstraction target was 10 weeks. This was accomplished after 10 weeks, as predicted, leading to a 60% reduction of backlogged cases. Conclusion: The delay of postdischarge chart abstraction was successfully shortened from 3.5 months to 1 month. We intend to maintain same-day abstraction efficiency without reaccumulating substantial backlog.

Key words: cardiac care, chart abstraction, National Cardiovascular Data Registry (NCDR), process improvement plan, Society of Thoracic Surgeons (STS) National Database

Introduction

Improving cardiac care has become a priority through the national reporting of several core measures. These measures serve to gauge the performance of an institution to the national average with respect to cardiac care and provide a benchmark that drives quality improvement. Institutions derive financial benefit from the Centers for Medicare and Medicaid Services (CMS) by adhering to several of these core measures recognized nationally. The St. Joseph’s Hospital/Marshfield Clinic Cardiac Database Registry submits data to both the National Cardiovascular Data Registry (NCDR) and to the Society of Thoracic Surgeons (STS) National Database. These data are obtained manually through retrospective abstraction of patient charts once they are released by the medical records department, typically 1 to 3 weeks after patients are discharged from the hospital. In addition to the national reporting of core measures, retrospective chart abstraction allows for the identification of missed opportunities of care that can be addressed by the providers involved. Upon identification of one of these missed opportunities of care, The Cardiac Database Registry sends a letter detailing the issue to the provider who was involved in the patient’s care while in the hospital. Some of these missed opportunities of care include medications not prescribed at time of discharge; the provider can address this issue by notifying the patient’s primary care provider, who can then prescribe the medication at a follow-up visit. Alternatively, the provider can notify the patient directly to provide a prescription. The time frame for such changes to be reflected in patient charts is narrow, since hospital policy prohibits any patient care clarifications from being made to the medical record more than 1 month after hospital discharge, as per CMS regulations. Therefore, delayed chart abstraction can potentially lead to financial loss from late adherence to core measures, and can also negatively impact patient care through the untimely notification to providers regarding missed opportunities of care.

Our institution was 3.5 months behind in postdischarge retrospective chart abstraction of cases being submitted to the CathPCI Registry within the NCDR. A process improvement initiative was created in order to reduce chart abstraction backlog to less than 1 month postdischarge, while keeping up with daily demand. In addition, this would allow for timely notification to providers regarding missed opportunities of care, allowing them to be resolved within the permissible time frame of the hospital and CMS for making patient care clarifications to the medical record. Due to financial limitations within the Cardiac Database Registry department, hiring new employees or using overtime was not possible. This led to the application of the lean approach in this initiative by using only existing resources. The process improvement plan was designed using the DMAIC (define, measure, analyze, improve, control) approach from Six Sigma methodology (Table 1).

Methods

The employees working in the Cardiac Database Registry had previous experience in health information data abstraction or research coordinator experience. They
were already assigned to specific registries for reporting data nationally. Cross training and equal division of abstraction was limited, since training for each specific module requires knowledge of specific definitions. Three employees were assigned to the CathPCI Registry submitting data to the NCDDR, which included diagnostic catheterizations and percutaneous coronary interventions (PCIs); while 1 employee was assigned to both the ACTION Registry–GWTG (Get with the Guidelines), also submitting data to the NCDDR, as well as the cardiac surgery registry submitting data to the STS National Database. The time required to abstract the various types of cases and to enter them into a local database were measured over the course of 1 year and averaged. Daily demand was calculated by determining the amount of data coming in for each registry per day. Next, daily capacity was calculated for 4 employees, assuming 6 hours per day dedicated to abstraction in an 8-hour work day, leaving 0.5 hours for lunch and 1.5 hours for other work, including ongoing quality assurance. Ten percent of all abstractions are reviewed by other employees within the Cardiac Database Registry for accuracy as per department policy. Daily demand was matched to daily capacity, and expectations were created for each employee’s daily work output. The time remaining per day was allocated toward reducing backlog. The abstraction goal was set as the expected backlogged case count that would remain at 1 month, based on the previous year’s data from the Cardiac Database Registry. Work output was measured for each employee, and progress was illustrated on a whiteboard.

### Table 1. Application of the DMAIC Approach Towards Developing a Process Improvement Plan

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Define</td>
<td>Retrospective case abstraction is occurring 3.5 months postdischarge.</td>
</tr>
<tr>
<td>Measure</td>
<td>Map out the current process of retrospective case abstraction.</td>
</tr>
<tr>
<td>Analyze</td>
<td>Identify methods to improve abstraction efficiency and reduce backlog.</td>
</tr>
<tr>
<td>Improve</td>
<td>Create new employee expectations, monitor performance, and display results.</td>
</tr>
<tr>
<td>Control</td>
<td>Maintain abstraction efficiency without reaccumulating backlog.</td>
</tr>
</tbody>
</table>

### Table 2. Daily Abstraction Demand

<table>
<thead>
<tr>
<th>Database for Submission</th>
<th>Cases per Day</th>
<th>Abstraction Time (min/case)</th>
<th>Total Minutes for All Cases</th>
<th>Total Hours for All Cases</th>
<th>Grouped Hours for All Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>CathPCI Registry</td>
<td>4</td>
<td>78</td>
<td>312</td>
<td>5.2</td>
<td>11.2</td>
</tr>
<tr>
<td>(percutaneous coronary interventions)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CathPCI Registry</td>
<td>8</td>
<td>45</td>
<td>360</td>
<td>6.0</td>
<td></td>
</tr>
<tr>
<td>(diagnostic catheterizations)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ACTION Registry–GWTG</td>
<td>2</td>
<td>42</td>
<td>84</td>
<td>1.4</td>
<td>5.9</td>
</tr>
<tr>
<td>STS National Database</td>
<td>2</td>
<td>135</td>
<td>270</td>
<td>4.5</td>
<td></td>
</tr>
<tr>
<td>Totals</td>
<td>16</td>
<td>—</td>
<td>1026</td>
<td>17.1</td>
<td>17.1</td>
</tr>
</tbody>
</table>

### Table 3. Daily Abstraction Capacity

<table>
<thead>
<tr>
<th>Database for Submission</th>
<th>No. of Employees</th>
<th>Available Hours (6 Hours per Employee)</th>
<th>Hours for Abstraction of Same-Day Cases</th>
<th>Remaining Same-Day Hours for Backlog Abstraction</th>
</tr>
</thead>
<tbody>
<tr>
<td>CathPCI Registry</td>
<td>3</td>
<td>18</td>
<td>11.2</td>
<td>6.8</td>
</tr>
<tr>
<td>(percutaneous coronary interventions)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CathPCI Registry</td>
<td>1</td>
<td>6</td>
<td>5.9</td>
<td>0.1</td>
</tr>
<tr>
<td>(diagnostic catheterizations)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ACTION Registry–GWTG</td>
<td>1</td>
<td>6</td>
<td>5.9</td>
<td>0.1</td>
</tr>
<tr>
<td>STS National Database</td>
<td>4</td>
<td>24</td>
<td>17.1</td>
<td>6.9</td>
</tr>
<tr>
<td>Totals</td>
<td>4</td>
<td>24</td>
<td>17.1</td>
<td>6.9</td>
</tr>
</tbody>
</table>

### Table 4. Utilization of Same-Day Hours for Backlogged Case Abstraction Based on 6.8 Hours/Day

<table>
<thead>
<tr>
<th>Database for Submission</th>
<th>Abstraction Time (min/case)</th>
<th>No. of Cases for Catch-up</th>
<th>Total Time (min)</th>
<th>Total Time (h)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CathPCI Registry</td>
<td>78</td>
<td>2</td>
<td>156</td>
<td>2.6</td>
</tr>
<tr>
<td>(percutaneous coronary interventions)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CathPCI Registry</td>
<td>45</td>
<td>6</td>
<td>270</td>
<td>4.5</td>
</tr>
<tr>
<td>(diagnostic catheterizations)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Totals</td>
<td>123</td>
<td>8</td>
<td>426</td>
<td>7.1</td>
</tr>
</tbody>
</table>
### Results

Daily abstraction demand was 16 cases requiring 17.1 hours (Table 2). Daily abstraction capacity was determined to be 24 hours for 4 employees, assuming 6 hours of effective abstraction time per day (Table 3). Same-day abstraction for cases being submitted to the ACTION Registry–GWTG and the STS National Database was found to be possible with only 1 employee assigned. Among the 3 employees assigned to abstract cases for submission to the CathPCI Registry, 6.8 hours per day remained for backlogged case abstraction. Based on this extra time available, feasible backlogged case abstraction was determined to be 8 cases per day, requiring 7.1 hours (Table 4).

Taking into account same-day and backlogged case abstraction requirements, 24 cases would need to be abstracted daily (Table 5). At the start of the initiative, there were 471 backlogged cases requiring submission to the CathPCI Registry. The abstraction target was set at 188 cases at 1 month postdischarge based on the previous year’s data (Table 6). For a standard 5-day work week, abstraction of the remaining backlogged cases for submission to the CathPCI Registry was predicted to be accomplished in approximately 10 weeks for PCIs and 6 weeks for diagnostic catheterizations (Table 6). Weekly performance of backlogged case abstraction revealed that backlogged case abstraction was completed within 10 weeks as predicted (Table 7). This led to an overall reduction of backlogged cases by 60%.

### Discussion

The success of our process improvement initiative was dependent on efficiently utilizing the time remaining after completing abstraction of same-day cases in order to steadily reduce backlog. Having employee work output measurement-driven has increased overall productivity in the department. After reaching the abstraction target, employees are now able to dedicate this extra time toward other projects. Because they receive hourly wages, higher productivity signifies increased employee value. The department has also been able to stay within budget, since the employees did not work overtime.

There was significant daily variation in the amount of data arriving for each registry. Because of this, the amount of backlogged case abstraction that could be completed varied from day to day. On lighter days, where same-day abstraction could be completed sooner, more backlogged charts were abstracted. Conversely, fewer backlogged charts were abstracted on busier days. In addition, the calculated abstraction times from Table 2 and Table 4 are averages and vary from case to case. These 2 sources of variation explain why 7.1 hours’ worth of backlogged case abstraction could be performed in the 6.8 hours remaining per day. Moreover, this initiative reduced the variation of total daily work output by utilizing the time beyond same-day case abstraction to reduce backlog. Variation in healthcare is known to increase costs, and reducing this variation is central to Six Sigma methodology.

---

**Table 5. Required Number of Cases to be Abstracted Daily to Reach Goal**

<table>
<thead>
<tr>
<th>Database for Submission</th>
<th>Same-Day Cases</th>
<th>Backlogged Cases/Day</th>
<th>Total Cases/Day</th>
</tr>
</thead>
<tbody>
<tr>
<td>CathPCI Registry (percutaneous coronary interventions)</td>
<td>4</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>CathPCI Registry (diagnostic catheterizations)</td>
<td>8</td>
<td>6</td>
<td>14</td>
</tr>
<tr>
<td>ACTION Registry–GWTG</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>STS National Database</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>16</strong></td>
<td><strong>8</strong></td>
<td><strong>24</strong></td>
</tr>
</tbody>
</table>

**Table 6. Predicted Time Required to Reach Abstraction Goal**

<table>
<thead>
<tr>
<th>Database for submission</th>
<th>Total Backlogged Cases</th>
<th>Backlogged Cases Remaining at 1 Month</th>
<th>Backlogged Cases to be Abstracted</th>
<th>Abstractions per Week</th>
<th>Time to Catch-up (weeks)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CathPCI Registry (percutaneous coronary interventions)</td>
<td>163</td>
<td>64</td>
<td>99</td>
<td>10</td>
<td>9.9</td>
</tr>
<tr>
<td>CathPCI Registry (diagnostic catheterizations)</td>
<td>308</td>
<td>124</td>
<td>184</td>
<td>30</td>
<td>6.1</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>471</strong></td>
<td><strong>188</strong></td>
<td><strong>283</strong></td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

**Table 7. Weekly Performance of Backlogged Case Abstraction**

<table>
<thead>
<tr>
<th>Week</th>
<th>% Backlogged Abstraction Completed</th>
<th>% Abstractions Remaining</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>98</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>83</td>
</tr>
<tr>
<td>3</td>
<td>24</td>
<td>76</td>
</tr>
<tr>
<td>4</td>
<td>42</td>
<td>58</td>
</tr>
<tr>
<td>5</td>
<td>45</td>
<td>55</td>
</tr>
<tr>
<td>6</td>
<td>62</td>
<td>38</td>
</tr>
<tr>
<td>7</td>
<td>77</td>
<td>23</td>
</tr>
<tr>
<td>8</td>
<td>86</td>
<td>13</td>
</tr>
<tr>
<td>9</td>
<td>97</td>
<td>3</td>
</tr>
<tr>
<td>10</td>
<td>100</td>
<td>0</td>
</tr>
</tbody>
</table>
This process improvement initiative had many strengths; however, its implementation met some challenges (Table 8). Initially, there was employee resistance when the concept of measurement-driven work output was introduced. This was mainly due to a lack of accountability involved in abstracting and entering data into a computer. Reinforcing the notion that patient care would improve with identifying and correcting missed opportunities of care helped to convince employees that this was a necessary change. We publicly displayed performance on a whiteboard in our department which was updated daily to provide incentive in meeting the increased work productivity expectations. This proved remarkably effective, in that healthy competition ensued between employees to further increase the number of charts abstracted. In addition, time off was not taken into account when planning this initiative. Fortunately, sick days and previously scheduled vacation time ultimately did not prolong the time to reach target. This is likely because there was some leeway in the predicted time to reduce backlogged cases being submitted to the CathPCI Registry (Table 6). Whereas reduction of backlogged cases to target had been predicted to take 10 weeks for the PCI section, the diagnostic catheterizations section was predicted to be completed much sooner, after only about 6 weeks. This difference of 4 weeks was offset by the time off taken by employees, causing both targets to be reached at the same time, after approximately 10 weeks.

We were successful in shortening the delay of retrospective postdischarge chart abstraction from 3.5 months to 1 month within 10 weeks, as predicted. This was accomplished without any new hires, overtime, or canceled vacations. Keys to the initiative’s success included making work output measurement-driven and utilizing extra time beyond same-day abstraction to reduce backlog. Providers within our institution are now happier to address missed opportunities of care in a timely fashion, and patient care clarifications can be made to the medical record up to 1 month postdischarge. Providers able to address missed opportunities of care sooner. Employee productivity increased, extra time can be allocated toward reduction of backlog and other projects. Providers able to address missed opportunities of care sooner.

<table>
<thead>
<tr>
<th>Strengths</th>
<th>Challenges</th>
</tr>
</thead>
<tbody>
<tr>
<td>Providers able to address missed opportunities of care sooner</td>
<td>Employees’ resistance to change</td>
</tr>
<tr>
<td>Patient care clarifications can be made to the medical record up to 1 month postdischarge</td>
<td>Introduction of work accountability</td>
</tr>
<tr>
<td>Increased reimbursement from CMS through higher adherence to core measures</td>
<td>Creating new expectations for employee work output</td>
</tr>
<tr>
<td>Higher employee productivity</td>
<td></td>
</tr>
<tr>
<td>Extra time can be allocated toward reduction of backlog and other projects</td>
<td></td>
</tr>
</tbody>
</table>

We were successful in shortening the delay of retrospective postdischarge chart abstraction from 3.5 months to 1 month within 10 weeks, as predicted. This was accomplished without any new hires, overtime, or canceled vacations. Keys to the initiative’s success included making work output measurement-driven and utilizing extra time beyond same-day abstraction to reduce backlog. Providers within our institution are now happier to address missed opportunities of care in a timely fashion, and patient care clarifications can be made to the medical record within the 1 month time frame as allowed by hospital policy and CMS. Successful chart abstraction within this time frame is dependent on the prompt release of patient charts soon after hospital discharge. In order to facilitate this, the Cardiac Database Registry maintains a good working relationship with the medical records department, which strives to release charts of patients who received cardiac care as soon as possible. Moving forward, we intend to maintain same-day abstraction efficiency without reaccumulating substantial backlog.

Table 8. Strengths and Challenge of Implementing the Process Improvement Plan

References
Updated and Expanded Study of Polycythemia Vera and Other Myeloproliferative Neoplasms in the Tri-County Area

J. M. Buchanich1; K. J. Mertz2; T. L. Washington3; J. N. Logue4; D. Marchetto5; P. I. Roda1; E. Irvin-Barnwell6

Abstract: Introduction: The results of a 2001–2005 polycythemia vera (PV) investigation in Eastern Pennsylvania revealed a disease cluster plus underreporting and false reporting to the Pennsylvania Cancer Registry (PCR). Purpose: The objectives of this study were 1) to assess PV reporting to the PCR in 2006–2009, 2) to determine whether a cancer cluster persisted, and 3) to determine whether other myeloproliferative neoplasms (MPNs), including essential thrombocytopenia (ET), were subject to similar reporting problems. Methods: Cases were identified from: 1) PCR records from the Tri-County, 2) reviewing billing records at Tri-County hematologist/oncologist offices, and 3) self-identification. An expert panel of physicians reviewed medical records and determined “true,” “false,” or “indeterminate” cases reported to the PCR. The analyses were conducted to determine sensitivity and positive predictive value (PPV) of case reporting to the PCR, estimate cancer incidence rates, and evaluate the presence of cancer clusters. Results: Of 290 cases identified, 90% were from the original PCR, 9% from billing records, and 1% from self-report. Fifty-five cases consented to participate, and medical records were obtained for 44. The expert panel determined that 45% were true cases, 32% were false cases, and 23% were indeterminate. PV had 100% (95% CI, 59–100) sensitivity, but only 47% PPV (95% CI, 20–70): ET had 78% (95% CI, 47–99) sensitivity and 100% PPV (95% CI, 59–100). Low participation and chart review rates led to rates with wide confidence intervals. We did not identify any PV cancer clusters, but we did identify a cluster of 9 ET cases in the Wilkes-Barre, Pennsylvania area. Conclusion: The current study was limited by the low response rate (22%) from MPN patients in the Tri-County area. This study identified 47% PPV for PV reporting and 100% PPV for ET.

Key words: epidemiologic methods, epidemiology, incidence, myeloproliferative disorders, polycythemia vera, registries

Background

Polycythemia vera (PV), a chronic hematologic malignancy involving an overproduction of red blood cells, belongs to a class of neoplasms classified by the World Health Organization (WHO) as myeloproliferative neoplasms (MPNs). All of the MPNs are hematopoietic stem cell disorders of common clonal heritage, characterized by bone marrow proliferation and peripheral blood erythrocytosis, thrombocytosis, or granulocytoses.1 In addition to PV, the MPNs include chronic myeloid leukemia (CML), essential thrombocytopenia (ET), primary myeloid fibrosis (PMF), and other related and unclassifiable MPNs, such as chronic neutrophilic leukemia.1 In 2005, a somatic point mutation in the JAK2 gene of hematopoietic cells was discovered; this mutation, JAK2V617F, is found in more than 90% of persons with PV and in approximately 50% of persons with ET and PMF.2 Factors leading to this acquired genetic mutation are unknown.

In 2004, physicians and residents in the Tamaqua area of eastern Pennsylvania became concerned about the diagnosis of PV in 4 persons living on the same street with nearby toxic waste sites.3 In 2005, the Pennsylvania Department of Health (PADOH) determined a higher incidence of PV in Luzerne and Schuylkill counties. Upon request from PADOH, the Agency for Toxic Substances and Disease Registry (ATSDR) assessed sensitivity and positive predictive value (PPV) of PV reporting to the Pennsylvania Cancer Registry (PCR) for Luzerne, Schuylkill, and Carbon counties. ATSDR used findings to estimate PV incidence rates from 2001 (when MPNs first became reportable) through 2005 in these 3 counties. The results of this evaluation indicated that inaccurate reporting of PV to the PCR led to PV risk estimates that were inflated over true values by 13% to 62%.2 The ATSDR study did identify a statistically significant cluster of PV cases near the intersection of the 3 counties. The incidence of PV in this cluster area was more than 4 times that of the entire Tri-County area. Several hazardous waste exposure sites were identified near the cluster area.3 In 2009, Congress funded ATSDR to continue this investigation. ATSDR is overseeing 18 projects related to this cluster with partners including the PADOH, the Pennsylvania Department of Environmental Protection, and various universities and private organizations.

The MPNs represent an inter-related series of diseases that may have a common origin, and the entire spectrum of these diseases has not yet been evaluated in the Tri-County area. The current study was designed as an update and expansion of the original ATSDR study to determine if...
1) sensitivity and PPV of PV reporting had improved in 2006–2009, 2) reporting of PMF, ET and related MPNs was complete and accurate in the Tri-County area for 2001–2009, and 3) rates of these related MPNs were elevated in the Tri-County area. The CML results are reported separately.

Methods

Case Ascertainment

The first phase of case ascertainment consisted of obtaining information on all cases reported to the PCR with an ID in 2007 or 2008. The study investigators assembled records received from medical providers and placed them in chronological order. Three expert panel members independently reviewed each case’s medical records. Expert panel members were instructed to review cases in 2 ways: 1) by applying conventional hematology practice standards at the time of diagnosis to determine the appropriateness or suitability of the diagnosis and 2) by classifying cases according to the 2008 WHO guidelines. Separate classification forms were developed for each disease. Expert panel members gave a determination for each case as “definitely” or “probably” a case (true cases), “possibly” a case (indeterminate), “definitely not” or “probably not” a case (false cases). At least 2 of the 3 opinions needed to be in agreement for true and false cases; if the members had different opinions or at least 2 members were not in agreement, the case was classified as indeterminate.

Data Collection

Potential cases were asked to participate in 3 phases of data collection: release of medical records related to their MPN diagnosis, a telephone survey, and a JAK2 mutation test (if one had not already been performed). Potential cases were mailed letters of introduction, consent forms, and releases for medical records (for review by the expert panel), and asked to return signed forms indicating their participation.

The study coordinator requested copies of outpatient and inpatient medical records relevant to the MPN diagnosis. The medical records submitted were reviewed. Incomplete medical record requests were also identified. The study coordinator telephoned the offices of medical providers and asked about the availability of medical records that appeared to be missing; multiple phone calls and repeated attempts to obtain records from noncompliant offices were made.

Patients not previously tested were asked to consent to JAK2 mutation testing. If they agreed, an appointment was scheduled at a local hospital for testing. A 10-cc blood sample was collected in an ethylenediaminetetraacetic acid (EDTA) blood collection tube and sent to the Division of Molecular Diagnostics, Department of Pathology, University of Pittsburgh Medical Center, for detection of the JAK2 mutation by allele-specific polymerase chain reaction.

Expert Panel Review

The 5-member expert panel consisted of 4 board-certified hematologist/oncologists and 1 family practitioner. The study investigators assembled records received from medical providers and placed them in chronological order. Three expert panel members independently reviewed each case’s medical records. Expert panel members were instructed to review cases in 2 ways: 1) by applying conventional hematology practice standards at the time of diagnosis to determine the appropriateness or suitability of the diagnosis and 2) by classifying cases according to the 2008 WHO guidelines. Separate classification forms were developed for each disease. Expert panel members gave a determination for each case as “definitely” or “probably” a case (true cases), “possibly” a case (indeterminate), “definitely not” or “probably not” a case (false cases). At least 2 of the 3 opinions needed to be in agreement for true and false cases; if the members had different opinions or at least 2 members were not in agreement, the case was classified as indeterminate.

Data Analysis

After cases were categorized as “true,” “indeterminate,” or “false,” we determined the sensitivity and PPV of the PCR. Sensitivity was calculated as the number of cases originally reported to the PCR divided by the total number of all true cases which includes those originally reported plus those found by reviewing billing records and self-report. PPV was calculated as the number of true cases originally reported to the PCR divided by the total number of cases, which included false positives, originally reported to the PCR. To adjust the PCR-reported incidence rates for sensitivity and PPV, we divided the Tri-County area incidence rate by the sensitivity then multiplied by the PPV. The 95% confidence intervals were calculated using the exact method.

To identify geographic subregions with an elevated incidence of an MPN, we included true cases and ZIP code- and census tract-level population counts from the US 2000 Census with ZIP code or census tract centroids (calculated from US Census Bureau shape files using ArcGIS Version 9 tools). We used SaTScan (Version 7.0.3), designed to analyze spatial, temporal, and space-time data using the corresponding scan statistics, and we used the Poisson-based model for spatial data, as well as the space-time permutation model. The discrete Poisson-based model considers the number of events in a geographical location as Poisson-distributed, based on the underlying population at risk. Under the null hypothesis, the expected number of cases in each area is proportional to its population size, or person years. The analysis is then conditioned on the total number of cases observed. The space-time permutation model uses only case data and scans for unusual occurrences in space and time simultaneously.
used the Poisson-based model for spatial data to identify a statistically significant primary geographic cluster of PV cases diagnosed in 2001–2005 in a region with a history of environmental contamination. This study was approved by the Institutional Review Boards of the University of Pittsburgh and the Pennsylvania Department of Health.

Results

Case Ascertainment

We identified a total of 290 potential MPN cases from the original PCR reports (n = 260), the enhanced casefinding (n = 25), and self-identification (n = 5). Overall, the original PCR data contributed approximately 90% of identified cases, with 42% reported as PV (Table 1). The PCR case-finding efforts identified an additional 25 cases, of which 56% (n = 14) were ET. Five cases self-identified to investigators: 4 PV cases and 1 ET case.

Case Participation

As shown in Figure 1, we determined a mailing address for 253, or 87%, of 290 cases. Of these, 58 consented (22%), 53 refused (21%), 2 (1%) were unable to consent, and 140 (55%) did not respond, despite numerous mailings and phone calls.

Over 90% of participants consenting to the study agreed to the interview (n = 54) and release of medical records (n = 55), but less than 20% consented to the blood draw (n = 10) (Figure 1). Of the 55 participants who agreed to the release of medical records, we were not able to obtain records for 11 (20%). Some physician offices (%) did not send records (n = 6) or required proof of executorship from the deceased cases’ estates (n = 5).

Expert Panel Review and Case Determination

Forty-four medical records were sent to expert panel members for review. Of these, 20 (45%) were determined to be true cases, 10 (23%) were indeterminant, and 14 (32%) were false (Figure 1). The expert panel members reached a unanimous opinion for 29 of the 44 cases reviewed (66%). PV had the lowest agreement rate (52%; 11/21 unanimous opinion) (data not shown).

Sensitivity and PPV of PCR Reporting

Case determination status by histology is provided in Table 2. ET had the highest percentage determined to be true cases (67%), followed by MPN/not otherwise specified (NOS) (60%), and PV and PMF (33%). Of the 44 evaluated cases, 36 were originally reported to the PCR, 3 were identified by review of billing records and 5 were self-identified. Seventeen of the 20 true cases were from the PCR; 2 were from billing records (1 ET and 1 PMF); and 1 from self-identification (ET).

Table 1. Case Ascertainment by Histology

<table>
<thead>
<tr>
<th>Histology</th>
<th>PCR</th>
<th>Casefinding</th>
<th>Self-Report</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
<td>%</td>
</tr>
<tr>
<td>PVa</td>
<td>110</td>
<td>91.7</td>
<td>6</td>
<td>5.0</td>
</tr>
<tr>
<td>ETb</td>
<td>84</td>
<td>84.8</td>
<td>14</td>
<td>14.1</td>
</tr>
<tr>
<td>PMFc</td>
<td>29</td>
<td>93.5</td>
<td>2</td>
<td>6.5</td>
</tr>
<tr>
<td>APMD</td>
<td>4</td>
<td>80.0</td>
<td>1</td>
<td>20.0</td>
</tr>
<tr>
<td>MPN/NOSd</td>
<td>33</td>
<td>94.3</td>
<td>2</td>
<td>5.7</td>
</tr>
<tr>
<td>Total</td>
<td>260</td>
<td>89.7</td>
<td>25</td>
<td>8.6</td>
</tr>
</tbody>
</table>

*Polycythemia vera, histology code 9950.
*Essential thrombocytopenia, histology code 9962.
*Primary myeloid fibrosis, histology code 9961.
*Acute panmyelosis with myelofibrosis, histology code 9931.
*Myeloproliferative neoplasm, not otherwise specified, histology code 9960.
Additional information about the indeterminate and false judgments is provided in Table 3. The majority of the indeterminate determinations (n = 7; 70%) were due to not enough information being provided in the medical records regarding criteria used for diagnosis. Of the false cases, many were determined to have a non-MPN diagnosis, primarily secondary polycythemia in patients diagnosed with PV. One self-reported case was deemed indeterminate but was probably a secondary polycythemia case. Three self-reported cases were deemed false: 1 did not have enough information in the medical records for the experts to make a determination, 1 was determined to be a JAK2+

Table 2. Expert Panel Determination by Histology

<table>
<thead>
<tr>
<th>Histology</th>
<th>Total</th>
<th>True Case</th>
<th>False Case</th>
<th>Indeterminate Case</th>
</tr>
</thead>
<tbody>
<tr>
<td>PV(^a)</td>
<td>23</td>
<td>7</td>
<td>11</td>
<td>5</td>
</tr>
<tr>
<td>ET(^b)</td>
<td>13</td>
<td>9</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>PMF(^c)</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>APMF(^d,)^(^e)</td>
<td>0</td>
<td>-</td>
<td>0</td>
<td>-</td>
</tr>
<tr>
<td>MPN/NOS(^f)</td>
<td>5</td>
<td>3</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Study Total</td>
<td>44</td>
<td>20</td>
<td>14</td>
<td>10</td>
</tr>
</tbody>
</table>

\(^a\) Polycythemia vera, histology code 9950.
\(^b\) Essential thrombocytopenia, histology code 9962.
\(^c\) Primary myeloid fibrosis, histology code 9961.
\(^d\) Acute panmyelosis with myelofibrosis, histology code 9931.
\(^e\) One APMF case identified through PCR enhanced case finding was reviewed for PMF diagnosis based upon investigator judgment and determined to be a true PMF case.
\(^f\) Myeloproliferative neoplasm, not otherwise specified, histology code 9960.

Table 3. Expert Panel Reason for Determination of “Indeterminate” or “False” Case Status

<table>
<thead>
<tr>
<th>EP Reason</th>
<th>Indeterminate</th>
<th>False</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>Not enough information/incomplete medical records</td>
<td>7</td>
<td>70.0</td>
<td>3</td>
</tr>
<tr>
<td>Non-MPN diagnosis</td>
<td>2</td>
<td>20.0</td>
<td>9</td>
</tr>
<tr>
<td>Other cancer</td>
<td>1</td>
<td>50.0</td>
<td>1</td>
</tr>
<tr>
<td>Secondary polycythemia</td>
<td>1</td>
<td>50.0</td>
<td>5</td>
</tr>
<tr>
<td>Other noncancer</td>
<td>0</td>
<td>0.0</td>
<td>3</td>
</tr>
<tr>
<td>Other MPN diagnosis</td>
<td>1</td>
<td>10.0</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>10</td>
<td>100.0</td>
<td>14</td>
</tr>
</tbody>
</table>

Table 4. Sensitivity and Positive Predictive Value of Original PCR Data by Histology

<table>
<thead>
<tr>
<th>Histology</th>
<th>“True” Cases Reported</th>
<th>“True” Cases Not Reported</th>
<th>“False” Cases Reported</th>
<th>Completeness (^a)</th>
<th>Accuracy (^d)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>%</td>
<td>%</td>
<td>%</td>
<td>95% CI (%)</td>
<td>%</td>
</tr>
<tr>
<td>PV(^e)</td>
<td>7</td>
<td>0</td>
<td>8</td>
<td>100.0</td>
<td>59.0 – 100.0</td>
</tr>
<tr>
<td>ET(^f)</td>
<td>7</td>
<td>2</td>
<td>0</td>
<td>77.8</td>
<td>47.3 – 99.7</td>
</tr>
<tr>
<td>PMF(^g,)^(^h)</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>0.0</td>
<td>0.0 – 97.5</td>
</tr>
<tr>
<td>MPN/NOS(^i)</td>
<td>3</td>
<td>0</td>
<td>1</td>
<td>100.0</td>
<td>29.2 – 100.0</td>
</tr>
<tr>
<td>Study Total</td>
<td>17</td>
<td>3</td>
<td>11</td>
<td>85.0</td>
<td>66.9 – 98.7</td>
</tr>
</tbody>
</table>

\(^a\) Cases that should have been included in the original PCR data set.
\(^b\) Cases incorrectly reported to the PCR.
\(^c\) True cases reported/(True cases reported + True cases not reported).
\(^d\) True cases reported/(True cases reported + False cases reported).
\(^e\) Polycythemia vera, histology code 9950.
\(^f\) Essential thrombocytopenia, histology code 9962.
\(^g\) Primary myeloid fibrosis, histology code 9961.
\(^h\) One APMF case identified through PCR case finding was reviewed for PMF diagnosis based upon investigator judgment and determined to be true PMF case.
\(^i\) Myeloproliferative neoplasm, not otherwise specified, histology code 9960.
non-MPN, and 1 was an “other MPN” diagnosis (PMF).

Table 4 shows the estimated sensitivity and PPV of the PCR. Sensitivity of the PCR was 85% (17 true PCR cases out of 20 true cases found in the study). The PCR data file included all true cases of PV. We estimated sensitivity of PV at 100% given that we did not find any additional true PV cases by searching billing records. The only true PMF case identified was originally reported to the PCR as APMF (code 9931) and after review by the expert panel was determined to be PMF. One ET cases was identified by the additional PCR casefinding efforts and 1 self-identified, giving ET a sensitivity of 78% (7/9). PV had the lowest PPV of 47% (7/15).

PV and Other MPN Incidence Rates

Shown in Table 5 are the original PCR incidence rates by histology, the study-determined sensitivity and PPV, and estimated incidence rates for comparison. As shown, our estimated ET incidence rate (2.3/100,000) is higher than the original PCR estimate (1.8/100,000). The other histologies have lower estimated incidence rates than those based on original case reporting. The estimated PV incidence rate showed the largest difference from the PCR-derived rate, at 5.3/100,000 compared to 2.5/100,000.

GIS Analysis

We performed SaTScan analyses by ZIP code and census tract for the 2 histologic groups with 5 or more true cases (PV and ET). Using the 7 true PV cases, no statistically significant clusters were identified in space or in space-time (the model adjusts for any purely spatial and temporal variation) at either the ZIP code or census tract level.

Using the 9 true ET cases, we identified a statistically significant cluster at the ZIP-code level when evaluated in space ($P < .05$), but not when using the space-time scan statistic ($I = .17$). The cluster includes 13 ZIP codes in the Wilkes-Barre, Pennsylvania area (Figure 2). The Poisson probability of finding 9 cases in this area, where 3.05 cases were expected, is .00029 ($P$-value).

Discussion

We found 89.5% sensitivity and 59% PPV of MPN reporting to the PCR as evaluated in this study, an expansion and update of an earlier ATSDR study in the Tri-County area of Carbon, Luzerne, and Schuylkill Counties, Pennsylvania.
In this study, the expert panel review confirmed an MPN in 54% of the evaluated cases, which was slightly higher than the original ATSDR investigation. However, only 47% of the PV cases evaluated in this update were determined to be true cases compared to 53% in the original ATSDR investigation. A companion study, conducted in a demographically similar 4-county region of Pennsylvania, found 82% sensitivity and 47% PPV for PV only in 2001–2009.

These findings indicate that MPNs remain very difficult to diagnose. The 2008 WHO guidelines could improve PPV of diagnoses; however, because our study period ended in 2009, the guidelines were not widely used or applied in this study. We also found that the inaccurate reporting was due to not distinguishing PV from other conditions, namely secondary polycythemia, and a lack either of JAK2 testing or documentation of such in the medical records. These results were surprising in view of the physician and hematologist education programs and extensive outreach that were conducted in the Tri-County area after completion of the ATSDR study, and the current widespread availability of the JAK2 test. Among cases evaluated by the expert panel, the PPV of PV reporting was only 47%, indicating that many false cases of PV are still being reported to the PCR. However, PV sensitivity was 100%, indicating that physician education and outreach efforts regarding the importance of PV reporting may have contributed to the increased reporting of PV in the Tri-County area. ET had better PPV than PV with a higher percentage of ET cases being confirmed as true cases.

Our estimated incidence rates are lower than rates calculated from the original PCR database reflect the reporting inaccuracies. The estimated PV incidence rate was 64% lower than the original rate, 2.5 (0.8–5.10) per 100,000 instead of 5.3, after correcting for sensitivity and PPV. According to the ATSDR study results, the annual incidence of confirmed PV was between 2.4 and 3.5 per 100,000 in Carbon, Luzerne, and Schuylkill Counties in 2001–2005. The wider range of values in this study reflects the variability associated with the findings based on the low response and review rate by the expert panel.

The original ATSDR study identified a statistically significant PV cluster in the Hazleton, Pennsylvania area with an incidence rate of 3.47; they found that the remainder of the Tri-County area had an incidence rate of 0.81 and the total area had an incidence rate 1.25. We did not identify any clusters of PV by ZIP code or census tract. We found a cluster of ET cases in the Wilkes-Barre area, based on 9 cases, which was statistically significant in space, but not in the space-time model at the ZIP-code level. Given that we are evaluating a 9-year time period, we place more emphasis on the space-time results, rather than those considering space only. Two of the ET cases were diagnosed in 1 census tract in a 2-year time period. Again, while this was statistically significant, it is difficult to determine the importance of such a small number of cases. Thirteen of 99 (13%) ET cases were evaluated by the expert panel. One lived in Carbon County, 1 lived in Schuylkill County and 11 lived in Luzerne County and all 9 true cases were Luzerne County residents. When all expert panel–evaluated ET cases (n = 13) were included in the cluster analyses, no statistically significant clusters were identified; similarly no clusters were identified using only the ET cases reported to the PCR (data not shown). The cluster identified here could be an artifact because all of the true cases resided in 1 county; ET cases in Luzerne may have been more willing to participate than cases in other counties. It may also represent a real increase of disease in Luzerne County. A more complete evaluation of ET might elucidate whether a cluster of ET persist in the Tri-County area.

This study was limited by a low incidence rate and a low response rate. The national incidence of MPNs has been estimated at 2.1 per 100,000. Additionally, only 26% of identified cases participated, although rates were slightly higher for some diseases, including PV. We attempted to include deceased cases in this study, which was not done in the original study. The participation rate among family members of deceased cases was significantly lower than the participation rate among living cases. Another reason for the low overall participation rate may be that the Tri-County area has been subject to numerous disease investigations during the past 20 years, in part owing to the high number of environmental contaminants in the region. Not only was the original ATSDR study conducted in the Tri-County area, but nearly a dozen other studies have been conducted in recent years, including some targeting the same cases who were asked to participate in this study. The Tri-County residents may be suffering from “study fatigue” and are no longer interested in cooperating with study investigators.

Despite the low response rate, our study provides important information on the sensitivity and PPV of MPN reporting to the PCR. We used press releases in Northeast Pennsylvania to recruit participants and performed extensive casefinding at hematology/oncology offices in and around the Tri-County area. We believe that these efforts completely and accurately captured the extent of the MPN cases in the Tri-County area for the time period of interest. Of the evaluated cases, we found that very few true MPN cases (n = 3) were missed in the original PCR data set. The PCR’s additional casefinding efforts identified 3 true cases, indicating that the use of billing information in outpatient settings may be an effective way for the PCR to gather case information from offices not reporting MPNs. The true self-identified ET case was from a facility with a hospital registrar in the Tri-County area. Outreach efforts regarding MPN reporting should potentially be expanded to hospital registrars, and not limited to physician offices.

Our updated and expanded study of MPNs in the Tri-County area identified continued low PPV for PV reporting, but better PPV for ET. These findings suggest the need for continuing physician and registrar education on diagnostic criteria, and increased use and interpretation of JAK2 testing for MPN diagnosis. Unlike the original study, we did not find any areas with a high occurrence of...
PV cases, although we did identify a cluster of ET cases (n = 9) in the Wilkes-Barre area in space, but not in space and time. The low case participation and case chart review rates may have led to sensitivity and PPV with wide confidence intervals and hampered our ability to identify statistically significant disease clustering.

References
Exclusion of Progressive Brain Disorders of Childhood for a Cerebral Palsy Monitoring System: A Public Health Perspective

Richard S. Olney, MD, MPH; Nancy S. Doernberg; Marshalyn Yeargin-Allsopp, MD

Abstract: Background: Cerebral palsy (CP) is defined by its nonprogressive features. Therefore, a standard definition and list of progressive disorders to exclude would be useful for CP monitoring and epidemiologic studies. Methods: We reviewed the literature on this topic to 1) develop selection criteria for CP monitoring and epidemiologic studies, 2) identify categories of disorders likely to include individual conditions that are progressive, and 3) ascertain information about the relative frequency and natural history of candidate disorders. Results: Based on 19 criteria that we developed, we ascertained a total of 104 progressive brain disorders of childhood, almost all of which were Mendelian disorders. Discussion: Our list is meant for CP surveillance programs and does not represent a complete catalog of progressive genetic conditions, nor is the list meant to comprehensively characterize disorders that might be mistaken for cerebral palsy. The criteria for progressive disorders that we developed could be applied by public health investigators in the future, as more children with very rare conditions are followed and new candidate disorders are identified.

Key words: cerebral palsy; neurodegenerative disease; population surveillance; public health

Introduction

Cerebral palsy (CP) is defined as “a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain.” Historically, the idea of nonprogression has always been a part of the definition of CP. Minear, who polled the membership of the American Academy of Cerebral Palsy in 1953, found that certain conditions were generally excluded: transient conditions, neoplasms, progressive disorders, and spinal cord disorders. Interestingly, these are still agreed-upon CP excludable conditions. Clinicians and researchers alike would seem to agree that “motor dysfunction which results from recognized progressive brain disorders is not considered CP.” While examples of such disorders have been published, drawn from cases found by population-based surveillance programs in Europe and Australia, we have not found a standard list of conditions that are considered progressive in registry management based on a literature review.

CP is a clinical condition, defined by history and physical findings, therefore diagnostic assessments are generally guided by clinical indications or suspicion of identifiable abnormalities. In 2004, the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society published a practice parameter that included an algorithm to assist with the diagnostic assessment of children with suspected CP. Whereas laboratory testing is not necessary to identify CP or its subtypes, studies of children with CP reviewed in the practice parameter found that a majority will have abnormalities on computed tomography (average, 77%, range, 62%-93%) or magnetic resonance imaging (average, 89%; range, 68%-100%). Metabolic or genetic testing has yielded abnormal results in children with CP less frequently, but such testing has been a consideration if the child has atypical features such as evidence of deterioration. Particularly for nonspastic clinical presentations associated with ataxia, dyskinesia, or hypotonia, confidence in CP categorization occurs only after assessments have been done for other possible neurological disorders (many of which are progressive). Therefore, surveillance personnel abstracting medical records and physicians caring for children with CP must be aware of specific neurological, genetic, and metabolic conditions that they might encounter.

The genesis of this literature review was a desire to construct a list of progressive conditions that most would agree are not CP, to assist nonphysician field staff reviewing and abstracting medical and education records in a community setting as part of the Autism and Developmental Disabilities Monitoring Network. This Network is a multisite, collaborative program funded by the Centers for Disease Control and Prevention to monitor the occurrence of developmental disabilities, including CP, in 8-year-old children across the United States. In this exploratory effort, our goal was to identify a list of brain disorders of childhood that by nature of their underlying pathophysiology and prognosis would not meet the nonprogressive component...
**Table 1. Public Health Surveillance Criteria for Progressive Brain Disorders of Childhood**

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
</table>
| 1 | A. For CP surveillance purposes, progressive disorders of childhood are those conditions causing progressive loss of motor skills (as opposed to those solely affecting memory and related dementia).  
B. The loss of motor skills must result from a recognized progressive brain disorder (as opposed to those solely of spinal, peripheral nerve or muscular origin). |
| 2 | A. For a condition to be considered a progressive disorder of childhood, the natural history of the condition should describe regression or a progressive or (neuro)degenerative course with onset during childhood. For CP surveillance purposes, “during childhood” is defined as ≤8 years old.  
B. If at least two references do not mention that the condition is progressive, then the condition is not progressive (eg, 18q-syndrome).  
C. If the typical age of onset for a progressive disorder is after age 8, then the condition is not considered a progressive disorder of childhood (eg, cerebral arteriopathy with subcortical infarcts and leukoencephalopathy – onset in midlife; earliest age in 20s).  
D. If fewer than 5 cases of a progressive disorder are reported in the literature, then the condition is not considered a progressive disorder of childhood for surveillance purposes. Rationale: a sufficient number of cases needs to be reported in the literature to obtain a general description of the natural history of the disorder. |
| 3 | A. If there are typical and atypical forms of a condition described in the literature, decide whether the condition is progressive based on what is true for the typical form of the disorder (eg, regression is seen in typical cases with Rett syndrome, but might not be a feature for atypical forms).  
B. If progression is a rare feature of a condition, do not consider the condition progressive (eg, craniometaphyseal dysplasia). Rationale: when the association is almost unheard of, do not exclude all potential children with CP who might coincidentally have the genetic condition.  
C. Conditions where progression during childhood is a possible but not universal feature (and progression is not a rare feature) will not be considered categorically progressive. Decisions about CP case status for individual children with these conditions should be made on a case by case basis through the review of the child’s medical history, motor findings and clinical course rather than the diagnosis per se.  
D. Progressive disorders that typically result in stillbirth or early mortality (before age 2) will not be included. Rationale: to be included in the monitoring program, the minimum age for CP diagnosis is age 2 years. In the unlikely event that a child with one of these disorders survives until age 8 and comes into the surveillance program, the decision about CP case status will be made on a case-by-case basis through the review of the child’s medical history, motor findings and clinical course. |
| 4 | A. For surveillance purposes, therapies to halt the progression of a condition will not be taken into account.  
B. Conditions that involve an accumulation of static cerebral lesions (eg, cerebrovascular complications of sickle cell disease) and predispose the child to repeated cerebral insults should not be considered progressive (“deterioration resulting from repeated insults is not the usual meaning of progressive”).  
C. Conditions where seizures are a feature (eg, tuberous sclerosis): do not take deterioration resulting directly from repeated seizures (“insults”) into account when deciding if the condition is progressive, as opposed to when the progressive effects of the underlying disorder cannot be separated from the associated seizures themselves, as in epileptic encephalopathies (eg, Dravet syndrome). |
| 5 | A. If the infantile/childhood form is progressive, then the condition is categorically progressive (eg, Krabbe disease or Alexander disease).  
B. Any condition with “adult onset” or “late onset” in the name is not considered a progressive disorder of childhood (eg, autosomal dominant late-onset leukoencephalopathy).  
C. If the condition is progressive during childhood and stabilizes during adulthood, then consider the condition progressive (eg, Sjogren-Larsson syndrome).  
D. If there are infantile and adult forms of a progressive condition, assume the child has the infantile form if the child shows neurologic signs during childhood. |
| 6 | A. Conditions described as acute are not considered progressive for surveillance purposes (eg, acute disseminated encephalomyelitis/ADEM).  
B. Conditions without clinical symptoms are not considered progressive for surveillance purposes (eg, extensive cerebral white matter abnormality without clinical symptoms). |
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<tr>
<th>MIM #</th>
<th>Disorder</th>
<th>Other Terms</th>
</tr>
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<td>Pantothenate kinase-associated neurodegeneration (PKAN)</td>
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Table 2, cont. Progressive Brain Disorders of Childhood for Public Health Surveillance

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<td>Sialuria, Finnish type</td>
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<td>Sjogren-Larsson syndrome (SLS)</td>
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<td>Zellweger syndrome (ZS)</td>
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*Multiple MIM (Mendelian Inheritance in Man) entries with same title root; all entries are progressive disorders; no entry has a commonly used eponym. Disorder is listed in this table with the MIM number for the most common MIM entry.

General term for progressive disorder; no MIM title; all subtypes are progressive disorders.

of the definition for CP. In this report, we present the methods for creating our list of progressive brain disorders of childhood and the table of such conditions identified to date.

**Methods**

*Criteria for Progressive Brain Disorders of Childhood*

As our first goal, we developed criteria for progressive disorders to apply in our literature review (Table 1). Since this activity was focused on a case definition for public health surveillance of CP, in particular for the Autism and Developmental Disabilities Monitoring Network, we concentrated on disorders with progressive features typically occurring by 8 years of age. By definition, we did not consider conditions that are purely myopathies, disorders only involving the spinal cord, or peripheral neuropathies (neuromuscular disorders), since the primary pathology in these conditions is not in the brain. Progressive features were defined primarily by loss of motor skills or milestones, although descriptions of disorders often more broadly described generalized regression, deteriorating clinical courses or neuropathological findings, or normal early development with subsequent developmental delay. If a disorder was clearly a neurodegenerative condition, we...
decided to list it for the purposes of exclusion from surveillance, even if some of the neurologic findings progressed and others did not. Another important feature that we considered was childhood mortality; lethality alone was not a criterion for progressiveness, since some genetic conditions known for mortality due to malformations or pathophysiologic processes outside of the central nervous system can have static or even improving neurologic manifestations.

The criteria took into account what is typical or described in the majority of children with disorders in question. The rationale for this principle was our belief that when neurologic deterioration is a rare feature, typical children with certain diagnoses who might have CP-like features for reasons unrelated to the disorder should not be excluded categorically. In practice, a limitation of applying this principle was the inadequate precision of literature quantifying the occurrence of CP-like features in rare genetic conditions. The issue of the effects of available therapies on natural history also is problematic, including the spectrum of interventions from diet and medications to enzyme replacement and stem-cell transplantation. Unfortunately, with our routine surveillance procedures, without a special study it is typically difficult to ascertain variables such as treatment regimens and timing of therapies that might be important in assessing the adequacy of treatment and its relationship to the clinical outcome of a particular child.9 For our list of progressive disorders, we did not review the core disorders on the Recommended Uniform Screening Panel for newborns in the United States,10 since the typical outcome for these conditions has changed because treatment is routinely instituted shortly after birth, thus preventing progressive features, eg, hypotonia and intellectual disability with congenital hypothyroidism. For other conditions with more potential variability in treatment in the general population, we did not consider the effects of such therapies on natural histories, eg, hematopoietic stem-cell transplantation in Krabbe disease. The rapid progress expected in the diagnosis and treatment of progressive disorders, with concomitant changes in newborn screening panels as well as clinical practice, is another caveat for the need to continuously update surveillance practices.

Selection Process for Categories of Candidate Conditions

After developing criteria for our literature review, the next step was to identify broad categories of disorders that were likely to include individual conditions that are progressive, such as leukodystrophies and lysosomal storage diseases. Ideas for these categories sometimes were generated by a particular disorder found in CP review articles or chapters,4,5,11-13 but we also attempted to identify potential categories through a search for progressive or degenerative disorders in published literature. Due to a lack of epidemiologic literature, we did not perform a formal meta-analysis but used standardized methods to review primarily expert literature (standard genetics textbooks, review articles, and selected online resources used in clinical practices and medical school courses). After a category of disorders was identified, we created a list of individual conditions within the category to review using overview chapters, indices, and summary tables in such literature.14-27 Occasionally, a new class was added after a query about a particular case by field staff.

Reviews of Candidate Conditions

Next, we searched for information about the natural history of candidate disorders in textbooks, review articles, and online catalogs14-27; if necessary, we evaluated primary sources of natural history data referenced therein or searched databases such as PubMed. Not uncommonly, natural history descriptions and neurologic manifestations were not mentioned in a particular review article or chapter, but only if 2 or more comprehensive sources lacked any information about a progressive course was the disorder left off our final list. A particularly important impetus for reviewing primary sources of information was when one source described a progressive neurologic feature but others did not. For example, the term progressive spasticity is used in connection with Weaver syndrome in a highly-cited textbook,18 but in a review by Opitz, Weaver, and Reynolds, this complication was described in only 1 child who also had spinal cord compression.28 Similarly, as noted in Table 1, we did not consider a condition to be a progressive disorder when deterioration tended to occur from repeated strokes or seizures per se, rather than events in the brain secondary to a neurodegenerative process.

Special Considerations for Conditions with High Rates of Fetal Death or Early Mortality

For surveillance purposes, we did not include conditions with high rates of fetal death or early mortality since the minimum age of CP diagnosis for inclusion in our monitoring program was 2 years (Table 1). For rare disorders, high rates of mortality are obviously problematic in assessing natural histories related to motor milestones, particularly when fetal or neonatal deaths are the typical outcomes. Occasionally children with such disorders will survive long enough to be ascertained by CP surveillance systems, and in fact children with some disorders described as lethal in older references are now treated surgically or with new medical interventions, and are gaining skills in special education settings. Our practice for such conditions is to make decisions about whether they should be excluded from CP surveillance on a case-by-case basis after they have been abstracted, rather than categorically labeling them as progressive disorders.

Special Considerations for Heterogeneous Conditions

We did not include groups of conditions with well-known clinical and genetic variability, such as mitochondrial neuromyopathies. Certain mitochondrial disorders were included if they resulted in a distinct syndromic phenotype that has a relatively well-defined natural history (eg, neuropathy, ataxia, and retinitis pigmentosa). Other mitochondrial disorders such as oxidative phosphorylation defects with specific electron transport complex pathology generally were not listed, since the nature of many of these conditions leads to heterogeneity of outcomes.
Some conditions such as Leigh syndrome are also heterogeneous but have a distinctive phenotype with progressive features generally included. There are also well-defined diagnostic criteria for such conditions with presumably less variability in community diagnoses. We therefore included such conditions on our list of progressive brain disorders. Some rarer conditions, such as pontocerebellar hypoplasia, have multiple genetic subtypes (with varying natural histories) that might not necessarily be evident to nonphysician field staff, and therefore would be considered on a case-by-case basis as described above.

Results

Table 1 includes all of the criteria we developed to define and select progressive brain disorders of childhood. Since we designed these 19 criteria for CP surveillance purposes, we qualified the overriding definition of a progressive disorder with that distinction (criteria 1A and 1B). The table includes some examples of disorders for which the selection process and special considerations were notably applicable (eg, criteria 2B, 3A, or 5A).

We have listed 104 disorders that we found that met our selection criteria in Table 2. Almost all of those itemized are Mendelian disorders, so we have also listed the Mendelian Inheritance in Man (MIM) numbers currently assigned to the disorders. The primary name usually corresponds to the main MIM title, but we have also listed other terms for clarity and for use by field staff.

Discussion

Many of these disorders that we identified for CP surveillance exclusion are quite rare, but together they represent a large number of affected children with individual metabolic and other genetic conditions that might be encountered by field staff. Our list does not represent a comprehensive catalog of progressive genetic conditions, nor does a condition’s absence from our list necessarily have clinical implications for a favorable prognosis. Readers should also note that some of these disorders would not be mistaken for CP by astute providers in many clinical settings; nevertheless, diagnoses of these progressive disorders should signal exclusion from ascertainment by surveillance program staff.

We found this review and compilation of conditions challenging for a number of reasons. First, there are few articles with a particular focus on these surveillance questions as they relate to CP. Hence, there is a need for this information but little to build upon. Secondly, the concept of a condition being slowly progressive is debated, but in the end, there is no consensus as to whether such a condition is considered progressive or not. Another challenge we found was with conditions where the clinical presentation varies considerably; eg, certain mitochondrial disorders. Without exact laboratory confirmation of type, how should conditions that fall within such a group be considered for possible exclusion as CP? If there is an atypical and typical form of the condition, we considered the clinical course of the typical form (eg, Rett syndrome). Any condition with a mean age of onset after age 8 or with “adult onset” or “late onset” in the name was not, for our purposes, considered a progressive disorder of childhood (eg, Friedreich ataxia).

Although this was a challenging undertaking, we think that it will have utility in surveillance and research as well as certain clinical settings. We will be applying the list in our own surveillance processes to determine its utility and validity, and in future work could analyze the practical use of this list to determine its value and to make updates. We also challenge others to critique our work and to expand upon it as more children with very rare disorders are followed and new candidate disorders are identified. By sharing our experience, we welcome others to consider the usefulness of defining what is and is not a progressive disorder and thereby extend the work we have started.

Attribution

We gratefully acknowledge Dr. Kim Van Naarden Braun and the anonymous peer reviewers for their helpful reviews and suggestions.

References


Cancers Coded as Tongue Not Otherwise Specified: Relevance to Surveillance of Human Papillomavirus–Related Cancers

Anthony P. Polednak, PhD; Cathryn Phillips, CTR

Abstract: Data from US population-based cancer registries have shown increasing incidence rates for cancer of the base of the tongue, interpreted as related to the epidemic of human papillomavirus (HPV) infection, but rates could be underestimated due to miscoding of some base of tongue cancers to tongue “not otherwise specified” (NOS). Tongue NOS was the most commonly coded subsite among incident (2000–2011) invasive cancers of the oral tongue (tongue excluding base of tongue and lingual tonsil which together comprise the posterior one-third of the tongue) in the 18 Surveillance, Epidemiology and End Results (SEER) Program registries combined and in the Connecticut SEER registry. All 173 cases of tongue NOS cancer in the Connecticut SEER registry diagnosed in selected years were reviewed. Only 5% were recoded to base of tongue, decreasing from over time from 8% to 2%, resulting in minimal impact on the incidence rate for base of tongue cancer in Connecticut. Most (76%) of the 173 tongue NOS cases were recoded to anterior two-thirds of tongue NOS, ruling out base of tongue as the actual site but resulting in underestimation of incidence rates for anterior two-thirds NOS in Connecticut. Similar studies are needed on tongue NOS cancers in other US cancer registries, along with studies on the HPV status of tumors at specific subsites of the oral tongue, to enhance surveillance and interpretation of trends in cancer incidence in relation to the HPV epidemic.

Key words: cancer registries, cancer surveillance, human papillomavirus (HPV), oral cancer, oropharyngeal cancer, tongue cancer

Introduction

Analyses of data from population-based cancer registries in the National Cancer Institute (NCI) Surveillance, Epidemiology and End Results (SEER) Program and/or the Center for Disease Control and Prevention (CDC) National Program of Cancer Registries (NPCR) have shown increasing incidence rates for cancers of base of tongue and certain other parts of the oropharynx for the US white population.1-4 These increases have been interpreted as largely related to the epidemic of human papillomavirus (HPV) infection in the population.1-5 Long-term increases in SEER incidence rates also have been reported for oral or mobile tongue cancers (ie, tongue other than base) in the oral cavity, which are weakly associated with HPV.6,7

Concerns have been raised about miscoding of base of the tongue cancers as other tongue subsites including tongue “not otherwise specified” (NOS),6-8 which could result in underestimation of the burden of HPV-related oropharyngeal cancers and trends in incidence of base of tongue cancers as well as projections of the future incidence rates for these cancers and possibly all HPV-related cancers.4

This study examined recent trends (2000–2011) in incidence rates for invasive tongue NOS cancer and other subsites of the tongue using data for the population-based Connecticut registry of the SEER Program and data for all 18 SEER registries combined. Potential miscoding of base of tongue as tongue NOS or other oral tongue subsites was addressed by manual review of cases coded as tongue NOS for selected years of diagnosis in the Connecticut registry. Implications of the findings were considered with regard to surveillance of trends in incidence rates for specific subsites of tongue cancers in relation to the HPV epidemic.

Methods

Incidence Rates for Tongue Cancer in Connecticut and All SEER Areas Combined

A de-identified research database available from the SEER Program website included cancers diagnosed in 2000–2011 in the 18 SEER registries, along with population estimates and analytic software (SEER*Stat version 8.1.5).9 The 18 registries include 9 with long-term incidence data (the states of Connecticut, Hawaii, Iowa, New Mexico, and Utah, and the metropolitan areas of Detroit; Atlanta; San Francisco-Oakland, California; and Seattle-Puget Sound Washington), and 9 others (Los Angeles, California; San Jose-Monterey, California; Alaska Native Registry; rural Georgia; Kentucky; Greater California; New Jersey; Louisiana; and Greater Georgia), together covering about 28% of the US population.9

The SEER site recode group “tongue” includes International Classification of Diseases for Oncology Edition 3 (ICD-O-3) site codes C019–029.9 Base of the tongue (C019) is part of the posterior one-third of the tongue (or pharyngeal tongue) which also includes lingual tonsil (C024). Site
C024 has been included with other tonsillar cancers (C09) and base of tongue, as HPV-related oropharyngeal cancers.\(^1\) The anterior two-thirds of the tongue, also known as oral or mobile tongue, was defined as including dorsal surface not otherwise specified (NOS) (C020), border or tip (C021), ventral surface NOS (C022), anterior two-thirds NOS (C023), overlapping lesion of tongue (C028), and tongue NOS (C029). Site C028 is sometimes included with HPV-related oropharynx\(^1\) while C029 could actually involve any part of the tongue. Selected were all invasive cancers of the tongue with ICD-O-3 Morphology code <9590 (M8000–9589, which excludes certain other cancers, mainly lymphomas).

Age-standardized incidence rates (ASIRs) and their 95% confidence intervals (CIs), for Connecticut and for the entire SEER-18 population, were analyzed (using 19 age groups from <1, and 1-4 through 80-84 years, plus 85+ years) using SEER*Stat.\(^9\) Statistical analysis of temporal trends (2000–2011) in ASIRs included the annual percent change (APC). APC was estimated as the slope of the line(s) obtained by fitting regression models (weighted least squares) to the natural logarithm of each annual rate, along with 95% confidence limits (CLs) on each APC, using SEER Joinpoint Regression Program (version 4.0.1, January 2013) to examine changes in the magnitude and/or direction of trends over the 12-year period of 2000–2011. For APCs, with zero as null hypothesis, \(P\) values were obtained from the joinpoint program (2-tailed tests); details on statistical methods used for these tests are available elsewhere.\(^2,10\)

Review of Cases Coded as Tongue NOS in the Connecticut Registry

For a sample of Connecticut cases coded as tongue NOS (C029), manual review of Connecticut registry records was conducted (by C.P., a certified tumor registrar) to determine if a more-specific site code could be assigned. Copies of full pathology reports are not routinely provided by 4 of the 32 Connecticut hospitals required to report cases to the Connecticut registry, or by some out-of-state sources, but detailed information abstracted from the pathology report is available in the synoptic summary routinely submitted to the Connecticut registry.

Manual review of all tongue NOS cases diagnosed since 2000 in the Connecticut registry was not feasible. Incidence data for 2012 were incomplete at the time of the study but were included to increase the number of recently diagnosed cases (2010–2012) available for review. The pre-2010 years of diagnosis selected included 2 years (2000–2001) before the introduction of American Joint Committee on Cancer (AJCC) Collaborative Stage (CS) coding that started with diagnoses in 2004, and the first 2 years of use of CS (2004–2005), in order to explore the potential impact of implementation of the CS staging schema on coding of subsite. The CS system includes coding of certain factors specific to subsites of the tongue,\(^11,12\) and this could have resulted in greater attention to coding of the specific subsite; alternatively, the increased workload involved with CS could have adversely affected the quality of subsite coding. The sample included all 49 registered cases coded as C029 diagnosed in 2000–2001, all 29 cases in 2004–2005, and all 95 cases in 2010–2012 (including 21 cases in 2010, 36 cases in 2011, and 38 cases in 2012), or a total of 173 cases, excluding 2 others ascertained by death certificate only (with unknown year of diagnosis). Statistical significance of differences in the distribution of recoded subsite by year of diagnosis category was tested by chi-square using Excel.

Results


For the Connecticut SEER area, the ASIR was higher for oral tongue (sites C020–023, C028, C029, and excluding C024) than for base of tongue (site C019) in 2000 and CIs did not overlap, but this was no longer true by 2011 because of a larger temporal increase for base of tongue than for oral tongue (Table 1). The most common subsite within the oral tongue in Connecticut was tongue NOS (C029), comprising 351 (38.8%) of all 811 (Table 1, Figure 1). The ASIR in Connecticut for site C029 was lower in 2000 (0.66, \(n = 24\)) than in 2011 (0.84, \(n = 36\)) (Table 1). There was an apparent temporary decline in the ASIR for C029 from 2001 through 2004, followed by an increase from 2005 to 2006 (Figure 1), but numbers of cases varied from only 15–26 per year and CIs on ASIRs overlapped (not shown); after 2006, there was no clear trend (Figure 1).

Data for other specific subsites of tongue in Connecticut are not tabulated because numbers were too small (<10 cases per year) for statistical analysis of trends.\(^11\)

For the SEER-18 areas combined, the ASIR for base of tongue was lower than that for oral tongue in 2000 but the ASIRs were similar by 2003 (Table 1, Figure 2). For tongue NOS, the most common site within the oral tongue (4,945/14,795 or 33.4%), the ASIR was higher in 2011 than in 2000 (Table 1) but the increase was mainly from 2005 to 2006, with no further increase (Figure 1). For specific subsites of oral tongue with sufficient numbers for analysis of temporal trends in SEER, the ASIR was higher for border of tongue than for anterior two-thirds NOS in 2000 but increased over time for anterior two-thirds NOS (APC = +2.6%) while decreasing for border of tongue (APC = –2.6%) (Table 1). The ASIRs for these 2 subsites had converged by 2004, with no clear trend for either subsite thereafter (Figure 2).

Review of Tongue NOS (C029) Cases in the Connecticut Registry

After manual review of the 95 cases diagnosed in 2010–2012 originally coded to C029, most (77%) were recoded as anterior two-thirds NOS (C023), 6% as ventral tongue (C022), and only 2% as base of tongue (C019), with 15% remaining as tongue NOS (Table 2); there was little difference in these proportions for 2010–2011 vs 2012 (for which data were preliminary). For those diagnosed in 2000–2001 and 2004–2005, 7%–8% of tongue NOS cases were recoded to base of tongue, 69%–76% to C023, and 12%–17% remained as C029 (Table 2). Comparing data for 2010–2012 vs all of the earlier years combined (due to small numbers per year), the overall distribution of recoded subsites was not statistically significantly different (\(P = .373\), but base
Table 1. Age-Standardized Incidence Rate per 100,000 for Invasive Tongue Cancers by Subsite Groups in the Connecticut (CT) SEER Registry vs all 18 SEER Registries Combined, for 2000 vs 2011, and Annual Percent Change (APC, %) in Rate from 2000–2011

<table>
<thead>
<tr>
<th>Area</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>APC (CL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Base of Tonguea</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CT</td>
<td>29</td>
<td>.80 (.53, 1.15)</td>
<td>79</td>
<td>1.75 (1.38, 2.20)</td>
<td>698</td>
<td>1.47 (1.36,1.58)</td>
<td>6.4 (2.8,10.1)*</td>
</tr>
<tr>
<td>SEER</td>
<td>897</td>
<td>1.21 (1.13,1.29)</td>
<td>1,592</td>
<td>1.66 (1.58,1.75)</td>
<td>14,942</td>
<td>1.48 (1.46,1.51)</td>
<td>3.0 (2.1,3.9)*</td>
</tr>
<tr>
<td>Oral Tongue, Totalb</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CT</td>
<td>58</td>
<td>1.59 (1.21,2.06)</td>
<td>74</td>
<td>1.75 (1.37, 2.21)</td>
<td>811</td>
<td>1.72 (1.61,1.85)</td>
<td>1.6 (-0.5,+3.7)</td>
</tr>
<tr>
<td>SEER</td>
<td>1,070</td>
<td>1.44 (1.36,1.53)</td>
<td>1,448</td>
<td>1.56 (1.48,1.65)</td>
<td>14,795</td>
<td>1.50 (1.47,1.52)</td>
<td>0.8 (-0.1,+1.7)</td>
</tr>
</tbody>
</table>

Subsites of Oral Tongue

2a. Tongue Not Otherwise Specified (NOS)

<table>
<thead>
<tr>
<th>Area</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>APC (CL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT</td>
<td>24</td>
<td>.66 (.42, .99)</td>
<td>36</td>
<td>.84 (.59, 1.18)</td>
<td>315</td>
<td>.67 (.60,.75)</td>
<td>4.1 (-1.5, +10.1)</td>
</tr>
<tr>
<td>SEER</td>
<td>326</td>
<td>.44 (.40,.49)</td>
<td>500</td>
<td>.54 (.49,.59)</td>
<td>4,945</td>
<td>.50 (.49,.52)</td>
<td>2.2 (0.7, 3.9)*</td>
</tr>
</tbody>
</table>

2b. Anterior Two-Thirds of Tongue NOSc

<table>
<thead>
<tr>
<th>Area</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>APC (CL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SEER</td>
<td>184</td>
<td>.25 (.21, .29)</td>
<td>328</td>
<td>.36 (.32,.40)</td>
<td>2,885</td>
<td>.29 (.28,.30)</td>
<td>2.6 (0.9, 4.2)*</td>
</tr>
</tbody>
</table>

2c. Border, Tipc

<table>
<thead>
<tr>
<th>Area</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>No.</th>
<th>Rate (CL)</th>
<th>APC (CL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SEER</td>
<td>319</td>
<td>.43 (.38,.48)</td>
<td>283</td>
<td>.31 (.27,.35)</td>
<td>3,581</td>
<td>.36 (.35,.38)</td>
<td>-2.6 (-3.8,-1.4)*</td>
</tr>
</tbody>
</table>

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*aInternational Classification of Diseases for Oncology Edition 3 (ICD-O-3) code C019.

*bIncludes dorsal (ICD-O3 C020), border or tip (C021), ventral including frenulum (C022), anterior 2/3 NOS (C023), overlapping lesion (C028), and tongue NOS (C029). Small numbers of cases of lingual tonsil cancer (C024) were excluded.

*cNumbers of incident cases for these sites in CT were too small for analysis of trends.

CL: Confidence limits (95%), lower and upper, which define the confidence interval.

SEER: Surveillance, Epidemiology, and End Results Program.

*Confidence limits on APC do not include zero.
Table 2. Invasive Cancers Originally Coded as Tongue Not Otherwise Specified (NOS), Diagnosed in Selected Years and Reported to the Connecticut Tumor Registry: Recoded Site after Manual Review of Records

<table>
<thead>
<tr>
<th></th>
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<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Base (C019)</td>
<td>4 8.2</td>
<td>2 6.9</td>
<td>2 2.1</td>
<td>8 4.6</td>
</tr>
<tr>
<td>Anterior 2/3 NOS (C023)</td>
<td>37 75.5</td>
<td>20 69</td>
<td>74 77.9</td>
<td>131 75.7</td>
</tr>
<tr>
<td>Tongue NOS (C029)</td>
<td>6 12.2</td>
<td>5 17.2</td>
<td>13 13.7</td>
<td>24 13.9</td>
</tr>
<tr>
<td>Other (C020, C022, C028)</td>
<td>2 4.1</td>
<td>2 6.9</td>
<td>6 6.3</td>
<td>10 5.8</td>
</tr>
<tr>
<td>Total</td>
<td>49 100.0</td>
<td>29 100.0</td>
<td>95 100.0</td>
<td>173 100.0</td>
</tr>
</tbody>
</table>

aData for 2012 are preliminary (incomplete), and not included on the SEER database used for other analyses in this report (see Table 1).

bIncludes dorsal (C020), ventral (C022) or overlapping lesion (C028). Excluded from the table were 2 cases ascertained by death certificate only.

The number of cases coded to C029 was lower for diagnoses in 2004–2005 than in other years (Figure 1).


Note: For results of statistical tests, see text.

(Table 2) would produce low and statistically unreliable rates; statistical analysis of trends would have required larger numbers (at least 10 cases per year).11

Discussion

For all 18 SEER registries combined, and for the Connecticut SEER registry, tongue NOS was the most common subsite within the oral tongue and the incidence rate increased from 2000–2006 (Table 1, Figure 1). A potential limitation of the analyses of trends is the lack of adjustment of rates for delayed reporting of cases to SEER registries, affecting mainly the most recent 2 years and cancers treated in nonhospital settings; however, the impact of adjustment for delayed reporting on trends in rates for oral cavity/pharynx cancers has been negligible in SEER.210

Incidence rates for base of tongue cancers increased from 2000–2011 for SEER registries and for the Connecticut registry alone (Table 1, Figure 2) but miscoding of some base of tongue cancers as tongue NOS could have affected the magnitude of the increase. In this study, however, after manual review of 173 cases originally coded as tongue NOS in the Connecticut registry, only 5% were recoded to base of tongue (Table 2), and this had a minimal impact on incidence rates for base of tongue cancer in Connecticut. Similar studies are needed in other central cancer registries.

In the Connecticut registry, the proportion of incident tongue NOS cases recoded to base of tongue declined from 7%–8% for diagnoses in 2000–2001 and 2004–2005 to 2% for 2010–2012 (Table 2) suggesting a recent improvement in coding of subsite of tongue. The decline was not consistent with the hypothesis of an immediate impact of the introduction of the CS system in 2004. Also, the number of incident cases originally coded to tongue NOS was temporarily low for 2004–2005 (Table 2), but had declined prior to 2004 (Figure 1). A limitation of the study is that 14% of tongue NOS cases reviewed could not be assigned a more specific site code (Table 2) and additional efforts such as follow-up with clinicians and pathologists would be needed to attempt to clarify the subsite of tongue involved.

Most (76%) of the sample of 173 cases in the Connecticut registry originally coded as tongue NOS were recoded to anterior two-thirds of tongue NOS (site C023) after review (Table 2), which ruled out base of tongue as the actual site but resulted in considerable underestimation of the incidence rate for site C023 in Connecticut. Although numbers of cases coded to C023 in Connecticut were too small for analysis of trends, the convergence in the ASIRs for site C023 and site C021 (border or tip of tongue) in the SEER-wide data (Table 1, Figure 2) may reflect a SEER ruling in calendar year 2004 that cases reported as simply “lateral” tongue should be coded as anterior two-thirds NOS.13

Cases reported as “lateral border” of tongue should have been coded to border of tongue, as described on lists of reportable neoplasms and in a SEER training module, but actual coding practices among central cancer registries should be assessed in future studies.

These findings on site C023 in the Connecticut SEER registry, and apparent temporal changes in coding practices in SEER-wide data, for certain subsites within the oral tongue may have limited impact on surveillance of HPV-related cancers, if the proportion of HPV-related cancers is low for each subsite of the oral tongue. Although HPV has been frequently detected in oropharyngeal cancers, including 70% of a sample of 213 invasive squamous cell cancers of base of tongue in a study using tissue repositories or other archived tissue in 7 US central cancer registries,10 detection in oral cavity cancers has been low but variable (about 0–25%). HPV DNA was detected in 27.8% of 54 cancers of the tongue excluding the base but including tongue NOS, ascertained from the metropolitan Detroit population-based registry, but the 95% CIs were 15.8% and 39.7%. Also, detection of HPV DNA by polymerase chain reaction (PCR) analysis of tumor tissue does not necessarily indicate a causal role of HPV. For tumor tissue HPV-positive by PCR, additional testing for persistent expression of HPV oncoproteins (mRNA) is regarded as more definitive evidence for a causal role. In what was reportedly the largest study of oral cavity tumors, with 409 consecutive cases from 4 North American hospitals, only 50% of those positive for HPV DNA were confirmed as positive for viral oncoprotein expression; the latter comprised only 24 (5.9%) of all 409. Six of these 24 were described as “tongue” but without specific-
only 1 (1.3%) was positive for HPV oncoprotein expression.\textsuperscript{20} In conclusion, the proportion of oral cavity cancers, including oral tongue, that are causally related to HPV may be small, although additional registry-based studies may be needed in archived tissues for larger samples of cancers at specific subsites of the oral tongue.\textsuperscript{a}

A potential source of registry-based information on HPV in oral tongue cancers is AJCC CS Site Specific Factor (SSF) 10, on the results of any HPV testing of tumor tissue; however, not all patients will have had their tumor tissue tested for HPV. AJCC has listed sites C020-029 as “discontinued” for SSF 10 for diagnoses since 2014 (as approved by SEER and NPCR),\textsuperscript{22} although never required by any standard-setting agency\textsuperscript{22} including NCI\textsuperscript{11} and CDC.\textsuperscript{23} Lindividual registries may continue collecting and storing data on discontinued SSFs.\textsuperscript{24} SEER registries have collected, for diagnoses since 2010, SSF 10 on HPV for base of tongue, tonsil and certain other oropharyngeal sites\textsuperscript{11,25} that have been classified as “HPV-related” or “HPV-associated” in surveillance reports; these data on SSF 10 have not been included in SEER-wide research databases.\textsuperscript{9,25}

Surveillance reports on trends in incidence rates have classified potentially HPV-related oropharyngeal cancers solely by cancer site codes, recognizing that not all cases are HPV-positive.\textsuperscript{1-3,5,26,27} Also, as noted above, oral tongue cancers (classified as “HPV-unrelated”) may involve a small proportion with HPV as a causal factor. Supporting the use of site codes for surveillance is the finding that miscoding of base of tongue to tongue NOS cancer was minimal in the Connecticut registry sample (Table 2), although studies are needed from other population-based registries. Such studies are important because base of tongue cancers comprised a substantial proportion (39%) of all incident invasive HPV-related oropharyngeal cancers in 2000-2011 defined by site codes\textsuperscript{1-3,5,25,27} and Morphology codes M8000-9589 using the SEER database\textsuperscript{9} (data not tabulated). HPV-associated oropharyngeal cancers comprised 78.2% of all incident invasive cancers at sites\textsuperscript{2b} classified as HPV-associated in men vs 11.6% in women (the majority being cervix), or 37.3% for both sexes combined, at age 15+ years in 2009 using data from the combined NPCR and SEER registries.\textsuperscript{2} Projected future numbers of HPV-positive oropharyngeal squamous cell carcinomas vs epithelial carcinomas of the cervix depend on such uncertainties as HPV vaccination coverage and vaccine efficacy for oral HPV infections.\textsuperscript{4}

Conclusions

Concerns have been raised regarding potential miscoding of base of the tongue cancers (strongly associated with HPV) to tongue NOS,\textsuperscript{2a} which was the most common subsite among oral tongue cancers diagnosed in 2000–2011 in SEER registries. After manual review of tongue NOS cases in the Connecticut SEER registry for selected years of diagnosis, however, only 5% were recoded to base of tongue, with minimal impact on incidence rates for base of the tongue in Connecticut. Most (76%) of the tongue NOS cases reviewed in Connecticut were recoded to anterior two-thirds of tongue NOS (site C023), which ruled out base of tongue, but had a substantial impact on the incidence of site C023 cancers in Connecticut. Reviews of tongue NOS cases are needed in other central cancer registries. Future studies should examine the reasons for the substantial proportion of oral tongue cancers that are coded to site C023 vs a more-specific subsite in SEER. Large registry-based studies are needed on the frequency of HPV detection in cancers at specific subsites of the oral tongue. All of these efforts may improve interpretation of trends in cancer incidence rates in relation to the HPV epidemic and efforts (such as HPV vaccination) to control the epidemic.\textsuperscript{16,27}

References


Decision Counseling and Participation in a Pancreas Cancer Registry

Ronald Myers, PhD; Harish Lavu, MD; Scott W. Keith, PhD; Heidi Kelly, MPH, BSN, RN; Nadine O’Rourke; James Cocroft, MA; Anna Quinn, MPH; Vishnu Potluri, MD; Charles J Yeo, MD

Abstract: Cancer registries play a vital role in research, as they provide important data that can be used to assess disease etiology and risk. Specialty registries can help to address the need for information on defined cancer types. However, achieving high rates of participation in such registries is problematic. We studied the impact of decision support on patient participation in a hospital-based pancreas cancer registry, the Jefferson Pancreas Tumor Registry (JPTR). In this study, we assembled a nonrandomized cohort of 40 patients, of whom 20 were exposed to the intervention and 20 were exposed to routine recruiting methods. Patients in the control group were invited to join the JPTR; while those in the intervention group were also invited to join the JPTR, and received decision support related to participation. Registry participation was assessed at 90 days. At baseline, patient gender, race, and stage of pancreatic cancer did not vary significantly between study groups. Overall, participation in the intervention group was significantly higher (P = 0.01) than in the control group (55% and 10%, respectively). In the intervention group, altruism was the major factor motivating patient participation, while patient concerns related to treatment recovery, registration time and complexity, and the confidentiality of registry data discouraged participation.

Key Words: registry, recruitment, cancer, pancreatic neoplasms, decision making

Introduction

Cancer registry data can help to advance research that could lead to improved pancreatic cancer prevention and control.1 There are different types of cancer registries, including Federal and state-mandated registries, collaborative registries, and voluntary hospital-based registries that have been developed to gather information on cancer cases in a standardized fashion, track cancer incidence, mortality, and survival, and to answer important research questions. In addition, cancer registries serve as a standardized system that can provide researchers with access to patient background information that may be used to learn about disease etiology and response to therapy. Cancer registries can also serve as resource for identifying patients and family members who may be invited to participate in a variety of research studies. Pancreatic cancer registries are important in these terms as they can facilitate much needed research on this devastating disease.

Pancreatic cancer is one of the most difficult types of cancer to cure, and survival is disappointingly low.2 Unfortunately, participation in pancreatic cancer registries is low. This situation limits research on specific exposures and inherited factors that contribute to risk and familial clustering of pancreatic cancer, and constrains efforts to identify patient characteristics that predict response to therapy.3 To advance research on pancreatic cancer prevention, early detection, and treatment, it is important to maximize participation in pancreas cancer registries.

The Jefferson Pancreas Tumor Registry (JPTR), established in 2008, is a hospital-based specialty registry maintained by the department of surgery at Thomas Jefferson University Hospital (TJUH) in Philadelphia, Pennsylvania. Recruitment to the registry routinely takes place for pancreatic cancer patients at TJUH both prior to and following initial surgery and also prior to discharge from the hospital. This in-person effort involves providing consented patients with print information about the JPTR, and encouraging consented patients to complete and return a survey questionnaire that records personal and family history, lifestyle, and environmental exposures. At the time of this study, Web-based registration in the JPTR was not an option. From February to December 2011, 175 pancreatic carcinoma patients underwent tumor resection at TJUH and were invited to participate in the JPTR. During this period, about 25% of patients treated at TJUH consented and completed the JPTR survey questionnaire. Participation in the JPTR involves signing an informed consent form and completing a survey questionnaire that includes items of sociodemographic background, personal and family medical history, and lifestyle.

In the current study, the research team sought to determine if decision counseling, a novel decision support intervention, had an effect on JPTR participation. During a typical decision counseling session, a trained counselor provides information about available options (eg, whether or not to participate in a pancreatic cancer registry), elicits pro and con factors that are likely to influence the patient’s
preference for the options, and clarifies the individual’s preferred option. The counselor shares session results with the patient and encourages performance of the preferred option. This approach has been used to aid patients in making decisions about cancer screening and genetic testing for cancer risk.\textsuperscript{13,15} We conducted a study, which was approved by the Jefferson Institutional Review Board, to determine the impact of decision counseling delivered by a trained health educator on JPTR participation.

**Methods**

Initially, the research team identified consecutively presenting patients who had a confirmed diagnosis of pancreatic carcinoma, underwent tumor resection at TJUH, were at least 18 years old, had not previously participated in the JPTR, and were scheduled to have a 1-month surgical follow-up consultation. At the follow-up visit, a practice clinical study coordinator met each identified patient, explained the decision-counseling study, obtained consent, and introduced consented patients to a trained study health educator at the conclusion of the surgical consultation. The health educator completed a decision counseling session (about 30 minutes) with the patient and accompanying significant others. Decision counseling sessions were conducted in the outpatient clinic or were completed by telephone after the visit in instances where time did not allow for the session in clinic.

Patients who underwent decision counseling were considered to be part of the intervention group. Retrospectively, we also identified 20 additional patients who had undergone resection for pancreatic cancer during a similar time period and had presented at the clinic for a 30-day follow-up visit. These patients were not approached by clinic staff for inclusion in the study due to clinic volume and thus were not exposed to decision counseling. These patients were defined as a nonrandomized control group. We assessed JPTR registration status (ie, completion of the JPTR survey) for both study groups at 90 days after their follow-up visit. Registration in the JPTR and other such registries is optional, which is not the case in federal and other mandated cancer registries.

**Decision Counseling**

Following standard methods reported elsewhere,\textsuperscript{4} the health educator initially reviewed a JPTR information page and registry survey questionnaire with each intervention group patient. Then, the health educator used a handheld computer software application (Decision Counseling Program) to help patients clarify whether they preferred option 1 (to participate in the registry) or option 2 (to not participate in the registry). Specifically, the health educator elicited decision factors (pros and cons) identified by the patient as being important in deciding on option 1 or 2; and asked patients to identify and rank the 3 top factors (primary, secondary, and tertiary) that were likely to influence their choice. Then, the patient assigned a weight to each factor corresponding to its level of importance, and compared the factors in terms of their relative importance. Decision importance weights were assigned to each pro

and con factor as follows: 1.0 (no importance), 1.3 (little bit of importance), 1.5 (some importance), 1.7 (much importance), 1.9 (very much importance), and 9.9 (overwhelming importance). The relative importance of each factor was also scored using a similar weighting scheme. The health educator entered all factor weights into the computer software program and generated a preference score for registry participation bounded between 0 and 1. A patient at equipoise regarding the decision would have a preference score of 0.5. Factors identified as favoring registry participation (pros) moved the preference score in a positive direction towards 1, while factors identified as not favoring participation (cons) moved the score in a negative direction towards 0. Finally, the health educator explained the preference result to the patient, verified the accuracy of the score, and produced a 1-page summary of the result, which was provided to the patient. At 90 days, a chart audit was conducted to determine patient JPTR participation status.

**Data Analysis**

We compared the intervention and control groups in terms of background characteristics including age, race, gender, and disease stage. Differences between the groups were evaluated by the Fisher exact test for comparing proportions, Student \( t \)-test for comparing means, or the nonparametric alternative Wilcoxon rank-sum test. Similarly, we assessed the associations of patient background characteristics and exposure to decision counseling with JPTR participation within 90 days after the follow-up visit. The significance level for all tests was set in advance at \( \alpha = .05 \). All statistical analyses were conducted using SAS version 9.3 (SAS Institute).

We reviewed decision factors elicited from each intervention group patient during the decision counseling session. The health educator (H.K.), study investigators (R.M., H.L.), and clinic staff (N.O.) classified these factors into pro and con categories, and calculated frequencies for each category. We then isolated the magnitude of influence each pro or con factor exerted on the individual’s preference score. The percentage of influence that a decision factor category had among all the participants was computed as the sum of factor-specific partial preference scores for all factors included in that factor category divided by the total sum of the absolute values of all factor-specific partial preference scores provided by the participants, multiplied by 100. Likewise, decision factor category influence percentages were computed for those who did not participate in the registry. These data allowed us to determine and display the relative influence of each factor category for registry participants and for nonparticipants.

**Results**

A total of 51 patients were initially identified as satisfying study eligibility criteria. Of this number, 42 returned to the surgical practice for a 30-day follow-up visit and were consented. Two were lost to follow up. Twenty consented patients received the decision counseling intervention (intervention group), while the remaining 20 patients (control group) did not.
Table 1 reports a descriptive summary of the patients’ background characteristics by study group. The intervention group and control group were similar in terms of their mean age (P = .59), gender (P = .99), and race (P = .69). Disease stage distributions were not significantly different between groups (P = .13). Table 2 shows the distributions of the patient background characteristics by JPTR participation status. Registry participation did not differ significantly by patient age (P = .29), gender (P = .31), race (P = .40), or disease stage (P = .17). However, patients who had decision counseling were significantly more likely to participate in the registry (P < .01). Table 3 presents decision factors reported by intervention group patients. Intervention group patients identified a total of 47 decision factors that were likely to influence whether they would join the JPTR. Factors that patients reported that would encourage them to join the registry (pro factors) included providing some benefit to others (altruism), gaining knowledge about their own health, having trust in researchers, and believing that providers/family members would want them to participate. Factors that would discourage them from joining the registry (con factors) included perceived registration time and complexity, concern about the confidentiality of information entered in the registry, and feeling overwhelmed by the treatment recovery process. Patients who joined the registry reported 15 pro and 10 con factors; while those who did not join the registry reported 12 pro and 10 con factors.

Figure 1 displays the relative aggregated influence of each reported decision factor category among registry participants and nonparticipants, respectively, as measured by factor-specific partial preference score percentages that reflected the influence of pro and con forces. Among registry participants, the overall influence of all pro factors (84%) was greater than the absolute value of con factor influence (17%). For nonparticipants, the overall influence of pro factors was lower (70%), while the absolute value of con factor influence was greater (30%) than was among registry participants. The strongest pro factor among both participants and nonparticipants was altruism, and this factor had more influence among participants than nonparticipants (77% and 59%, respectively). Patient concerns about time and complexity related to participating in the JPTR were expressed by both participants and nonparticipants. Interestingly, the influence of this con factor was greater among participants compared to nonparticipants (13% and 4%, respectively). Participants and nonparticipants also expressed concerns related to coping with the treatment recovery process, but this con factor had much more influence among nonparticipants (16%) than participants (3%). The desire to gain knowledge and having trust in researchers were pro factors mentioned exclusively by registry participants (5% and 2%, respectively). Perceived social support for joining the registry (8%) and concerns about the confidentiality of registry data (10%) were reported only by nonparticipants.

Table 1. Background Characteristics of Patients by Study Group

<table>
<thead>
<tr>
<th></th>
<th>Intervention Group (n = 20)</th>
<th>Control Group (n = 20)</th>
<th>P-Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mean (SD)</td>
<td>66.2 (11.8)</td>
<td>64.1 (12.8)</td>
<td>.59</td>
</tr>
<tr>
<td>Gender, n (%)</td>
<td></td>
<td></td>
<td>.99</td>
</tr>
<tr>
<td>Women</td>
<td>10 (47.6)</td>
<td>11 (52.4)</td>
<td></td>
</tr>
<tr>
<td>Men</td>
<td>10 (47.6)</td>
<td>11 (52.4)</td>
<td></td>
</tr>
<tr>
<td>Race, n (%)</td>
<td></td>
<td></td>
<td>.69</td>
</tr>
<tr>
<td>Whites</td>
<td>15 (46.9)</td>
<td>17 (53.1)</td>
<td></td>
</tr>
<tr>
<td>Blacks</td>
<td>5 (62.5)</td>
<td>3 (37.5)</td>
<td></td>
</tr>
<tr>
<td>Disease Stage, n (%)</td>
<td></td>
<td></td>
<td>.13</td>
</tr>
<tr>
<td>Pancreatic neoplasm</td>
<td>3 (60.0)</td>
<td>2 (40.0)</td>
<td></td>
</tr>
<tr>
<td>T1</td>
<td>1 (100.0)</td>
<td>0 (0.0)</td>
<td></td>
</tr>
<tr>
<td>T2</td>
<td>4 (66.7)</td>
<td>2 (33.3)</td>
<td></td>
</tr>
<tr>
<td>T3</td>
<td>12 (54.5)</td>
<td>11 (46.5)</td>
<td></td>
</tr>
<tr>
<td>Metastatic disease</td>
<td>0 (0.0)</td>
<td>5 (100.0)</td>
<td></td>
</tr>
</tbody>
</table>

Table 2. Factors Associated with Patient Participation in the Registry

<table>
<thead>
<tr>
<th></th>
<th>Participant (n = 13)</th>
<th>Nonparticipant (n = 27)</th>
<th>P-Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mean (SD)</td>
<td>67.2 (14.3)</td>
<td>64.1 (11.3)</td>
<td>.29</td>
</tr>
<tr>
<td>Gender, n (%)</td>
<td></td>
<td></td>
<td>.31</td>
</tr>
<tr>
<td>Women</td>
<td>5 (23.8)</td>
<td>16 (76.2)</td>
<td></td>
</tr>
<tr>
<td>Men</td>
<td>8 (61.5)</td>
<td>11 (50.0)</td>
<td></td>
</tr>
<tr>
<td>Race, n (%)</td>
<td></td>
<td></td>
<td>.4</td>
</tr>
<tr>
<td>Whites</td>
<td>9 (28.1)</td>
<td>23 (71.9)</td>
<td></td>
</tr>
<tr>
<td>Blacks</td>
<td>4 (50.0)</td>
<td>4 (50.0)</td>
<td></td>
</tr>
<tr>
<td>Disease Stage, n (%)</td>
<td></td>
<td></td>
<td>.17</td>
</tr>
<tr>
<td>Pancreatic neoplasm</td>
<td>2 (40.0)</td>
<td>3 (60.0)</td>
<td></td>
</tr>
<tr>
<td>T1</td>
<td>0 (0.0)</td>
<td>1 (100.0)</td>
<td></td>
</tr>
<tr>
<td>T2</td>
<td>4 (66.7)</td>
<td>2 (33.3)</td>
<td></td>
</tr>
<tr>
<td>T3</td>
<td>7 (30.4)</td>
<td>16 (69.6)</td>
<td></td>
</tr>
<tr>
<td>Metastatic disease</td>
<td>0 (0.0)</td>
<td>5 (100.0)</td>
<td></td>
</tr>
<tr>
<td>Study Group, n (%)</td>
<td></td>
<td></td>
<td>&lt; .01</td>
</tr>
<tr>
<td>Intervention</td>
<td>11 (55.0)</td>
<td>9 (45.0)</td>
<td></td>
</tr>
<tr>
<td>Control</td>
<td>2 (10.0)</td>
<td>18 (90.0)</td>
<td></td>
</tr>
</tbody>
</table>

Discussion

The current study is the first to evaluate the impact of decision counseling on participation in a specialty cancer registry. We found that patients exposed to decision counseling were significantly more likely to participate in the registry than those who were not. Our findings are consistent with other reports showing a stronger, positive impact of active engagement registry recruitment strategies compared to passive engagement strategies. In a 2004 study that focused on recruiting patients from cancer risk assessment clinics to join a national hereditary cancer registry, Freibell et al sought to find that direct recruiter contact had substantially greater impact on recruitment than a brochure
Table 3. Distribution of Primary, Secondary, and Tertiary Decision Factors among Patients in the Intervention Group by Participation Status (n = 20)

<table>
<thead>
<tr>
<th>Participation Status/Pro and Con Factors</th>
<th>Decision Factor</th>
<th>Primary</th>
<th>Secondary</th>
<th>Tertiary</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Participants (n = 11)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pro</td>
<td>Provide benefit to others</td>
<td>11</td>
<td>0</td>
<td>1</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td>Gain knowledge about health</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Trust in researchers</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Social support</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Con</td>
<td>Perceived time and complexity of registration</td>
<td>0</td>
<td>7</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>Concern about confidentiality</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Feeling overwhelmed by treatment recovery</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>11</td>
<td>11</td>
<td>3</td>
<td>25</td>
</tr>
<tr>
<td>Nonparticipants (n = 9)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pro</td>
<td>Provide benefit to others</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>Gain knowledge about health</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Trust in researchers</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Social support</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Con</td>
<td>Perceived time and complexity of registration</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Concern about confidentiality</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Feeling overwhelmed by treatment recovery</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>9</td>
<td>8</td>
<td>5</td>
<td>22</td>
</tr>
</tbody>
</table>

Figure 1. Distribution of Decision Factor Influence among Intervention Group Registry Participants (n = 11) and Nonparticipants (n = 9)

mailing (67% and 16%, respectively). In a study that focused on recruiting African American patients to a national cancer genetics registry, Skinner et al.\(^8\) reported that the combined mailing of print education materials and follow-up telephone contact resulted in a high level of enrollment (68%). Ramirez et al.\(^9\) also found that the combination of mail and telephone contact generated higher cancer registry participation (37%) than either mail or telephone alone (31% and 25%, respectively). Registration intervention studies reported by Patterson et al.\(^10\) and Wenzel et al.\(^11\) add to the literature supporting more active engagement in registry recruitment. A review of factors identified in the decision counseling sessions revealed that registry participants and
nonparticipants were both motivated by a strong sense of altruism, the belief that participating in the JPTR might benefit others. Both participants and nonparticipants also identified concerns about the complexity of the registration process and feeling overwhelmed by the process of recovering from surgery as obstacles to joining the registry. Closer inspection of identified decision factors indicates that altruism was the most influential force among JPTR participants, however. While also important among registry nonparticipants, this important pro factor seems to have been counterbalanced by con factors that included concerns related to treatment recovery, the confidentiality of registry information, and the time and complexity of registration.

This study has several limitations that may limit generalizability. First, the study was conducted at 1 site, with a small number of pancreatic cancer patients. Second, the study was not a randomized controlled trial. While there were no significant distributional differences between the study groups with respect to age, race, gender, or disease stage, it is possible the groups differed in other ways that could have biased the results. Third, among intervention group patients, 5 had decision counseling in the clinic and 15 completed the session over the telephone (after follow-up). While there was no statistically significant difference in registry participation rates between those who received clinic vs telephone decision counseling, registry participation rates were substantially higher among those who were counseled in the clinic compared to those who were counseled by telephone (80% vs 47%, respectively). This observation suggests that in-person counseling may be a more effective than telephone contact. Finally, online recruitment methods are being developed and deployed to facilitate recruitment to cancer registries. This technology was not integrated into the JPTR at the time the study was conducted, however. Thus, we were not able to determine the feasibility or impact of embedding the Decision Counseling Program in an overall Web-based recruitment process.

**Conclusion**

Findings from the current study suggest that decision counseling boosted participation in a specialty cancer registry. We also observed that patients treated for late-stage disease may have special needs and concerns that could restrict participation. To date, the JPTR includes more than 400 participants, and the registration effort is ongoing. Further research is needed to assess the feasibility of integrating decision counseling into ongoing efforts to facilitate patient participation in this and other types of cancer registries. In addition, randomized trials are needed to more definitively evaluate the impact of decision counseling on the recruitment to cancer registries. Close inspection of factors influencing registry participation suggests that particular attention should be devoted to identifying and addressing the needs of patients who find participation to be challenging due to concerns related to recovery from treatment and confidentiality.

**References**

Original Article

What Increased Registry Outreach May Mean for Cutaneous Melanoma Surveillance: Impact of Changes in Iowa

Mary E. Charlton, PhD; Kiran Sapkota, MS, MPH; Melody J. Eide, MD, MPH; Daniel B. Olson, MS; Kathleen McKeen; Charles E. Platz, MD; Jennifer A. Schlichting, PhD; Charles F. Lynch, MD, PhD;

Abstract: Background: Cutaneous melanoma (CM) is underreported to cancer registries, in part due to insufficient reporting in the nonhospital setting. The objective of this study was to better understand the impact of dermatologist and private pathology laboratory reporting on CM rates. Methods: We examined the impact of targeted casefinding in private pathology laboratories and dermatology offices by the State Health Registry of Iowa (SHRI) on CM incidence, as well as the characteristics of nonhospital reported cases. Results: Over the 39-year period (1973–2011), 22,541 cases of CM were captured by the SHRI; 16,183 (72%) were invasive melanoma cases and 6,358 (28%) were in situ cases. The incidence of invasive melanoma increased 3.6 fold between the time periods of 1973-1975 and 2009-2011 (6.6 vs. 24 per 100,000 person-years, respectively). If case reporting from private pathology laboratories and dermatology offices was not conducted, the 2009–2011 invasive CM rate would have decreased to 19.1. The ratio of invasive to in situ cases declined from 8:1 from 1973–1987 to less 2:1 from 2007–2011. Age at diagnosis also significantly increased across time periods, while the proportion of females declined. From 2007–2011, the majority (55%) of nonhospital cases were in situ, and 90% of the invasive cases were localized. A higher percentage of urban residents were attributed to nonhospital-based reporting sources compared to hospital-based sources (57% vs 45%, P < .0001) Conclusions: Electronic health records and incentivized Meaningful Use for reporting may provide an efficient method for nonhospital based providers to easily and accurately report CM cases to registries.

Key words: cancer registries, cancer reporting, cutaneous melanoma, dermatology office, incidence rates, private pathology laboratory

Introduction

The incidence rate of cutaneous melanomas (CM) in the United States has increased significantly over the past 20 years, with several factors likely contributing to the increase, including changes in ultraviolet (UV) light exposure as well as increased public awareness and clinical surveillance. Changes in cancer reporting are another potentially important contributor to CM trends. Every US state requires cancer be reported to the state’s central cancer registry. In the past, collecting and reporting of cases was done almost exclusively in hospital facilities. However, cancers such as early stage CM are increasingly diagnosed and treated outside of the hospital setting. Furthermore, the increase in nonhospital affiliated independent laboratories, including national private laboratories, has provided more options for clinicians to have their pathology specimens evaluated, thereby circumventing traditional hospital laboratory-based reporting.

Several studies have demonstrated underreporting of CM in the United States, with estimates from different registries indicating between 10% and 20% of CM cases were missed from the 1980s through the early 2000s, and as high as 30% to 40% of cases were missed in parts of California in 2005-2006. Furthermore, among 104 dermatologists surveyed during the 2010 American Academy of Dermatology meeting, 50% were unaware they are required to report CM and 56% did not actively report to a cancer registry.

We examined the impact of recent efforts to engage nonhospital-based providers, namely private pathology laboratories and dermatologists, in reporting incident in situ and invasive CM to the State Health Registry of Iowa (SHRI). The aims of this study were to better understand the impact of these reporting sources on CM incidence rates as well as the characteristics of cases reported from nonhospital sources.

Methods

Study Population

All in situ and invasive CM diagnosed in Iowa residents between 1973 and 2011 and captured by the SHRI were included in the study. Data were extracted from the SHRI database. This project was granted human subject exemption status by the University of Iowa Institutional Review Board.

Reporting Sources

While cases are often reported by more than one source, they can only be attributed to a single source based on a hierarchy defined by the Surveillance, Epidemiology,
and End Results (SEER) Program Coding and Staging Manual, which considers hospital inpatient to be the best information, followed by radiation or medical oncology treatment centers, physician’s office, pathology laboratory only, nursing home and finally autopsy or death certificate only.\textsuperscript{14} Reporting sources were collapsed into the following categories for analysis: 1) hospital facilities, which include hospital-based pathology laboratories, outpatient facility-based clinics, ambulatory surgery centers and free-standing radiation treatment or medical oncology centers, 2) private pathology laboratories, which are independent from any hospital facility, and 3) dermatology offices not owned by or located within a hospital facility.

**Time Periods**

In order to display trends with sufficient granularity over the 39-year study period, incidence rates were calculated in thirteen 3-year increments. In addition, 3 distinct time periods were defined to reflect changes in SHRI operations related to reporting of CM cases: 1) 1973–1987, SHRI focused almost exclusively on casefinding in hospital facilities; 2) 1988–2006, intensified SHRI efforts to acquire records from local and national private laboratories; 3) 2007–2011 (most recent reporting year), SHRI began reaching out to dermatology clinics across Iowa in order to acquire case reporting for patients who might not be receiving facility-based treatment for their CM, with many of the surgical specimens sent to out-of-state laboratories.

**Demographic and Clinical Variables**

Information on age, gender, race, stage, skin subsites, morphology, and rurality were extracted for all CM cases from the SHRI data. Age was evaluated both as a continuous variable and in categories of <40, 40–59, 60–79 and 80+ years of age. Given the low proportion of nonwhite populations in Iowa and the much higher incidence of CM in the white population, race was categorized as white vs other (ie, nonwhite and unknown). As this study period spanned 39 years, SEER Historic Stage was used to describe stage among invasive CM cases in terms of localized, regional, and distant disease.\textsuperscript{15} Skin subsites (ie, International Classification of Diseases for Oncology-3 site codes C440–C449) were collapsed into the categories of head and neck, trunk, upper extremities, lower extremities, and skin not otherwise specified (NOS).\textsuperscript{16} Morphology was categorized as superficial spreading, nodular, lentigous, amelanotic, epithelioid, regressing, and balloon cell melanomas, and malignant melanoma in giant pigmented nevus or in precancerous melanosis or malignant blue nevus).\textsuperscript{16}

Rurality was based on Rural-Urban Commuting Areas (RUCA), which classify US ZIP codes based on population density, urbanization, and daily commuting into 30 mutually exclusive categories. These were then collapsed into 4 previously defined categories: urban areas, large rural towns, small rural towns, and isolated small towns.\textsuperscript{17}

**Data Analysis**

In situ and invasive CM frequency counts and incidence rates age-adjusted to the 2000 US standard population were calculated in SEER\textsuperscript{®}Stat (version 8.1.5). Differences between in situ and invasive CM were assessed using t-tests for continuous variables and chi-square tests for categorical variables. Differences in in situ and invasive CM across the 3 time periods were assessed using analysis of variance (ANOVA) for continuous variables and chi-square tests for categorical variables. Analyses were conducted using SAS software (version 9.3, SAS Institute).

**Results**

From 1973–2011, 16,183 (72%) invasive and 6,358 (28%) in situ CMs were recorded at SHRI, with the incidence of both in situ and invasive CM increased markedly over the 39-year time period (Figure 1). Figure 1 also demonstrates if case reporting from private pathology laboratories and dermatology offices was not conducted, the 2009–2011 invasive CM incidence rate (IR) would have been underestimated as 19.1 per 100,000 person-years, which is 4.9 cases per 100,000 lower (20% reduction) compared to the rate which includes additional case reporting from private pathology laboratories and dermatology/private physician offices (IR 24.0 per 100,000). Likewise, incidence of in situ CM would have been 6.9 per 100,000 person-years if based on hospital facility reporting only, which is 5.5 cases per 100,000 lower (44% reduction) than the incidence based on all reporting sources (IR 12.4 per 100,000).

Table 1 shows case characteristics across the 3 time periods (1973–1987, 1988–2006, and 2007–2011). There were statistically significant changes in CM case characteristics across time (all $P \leq .0003$). Most notably, the ratio of invasive to in situ cases declined dramatically from 8:1 from 1973–1987 to less than 2:1 from 2007–2011. Age at diagnosis significantly increased across time periods, with the proportion of those 80 years of age and older diagnosed with CM nearly doubling from 11% from 1973–1987 to 19% from 2007–2011. The proportion of females diagnosed declined
**Table 1. Characteristics of Cases of Cutaneous Melanoma Ascertained in Iowa by Time Period**

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong> Mean (SD)</td>
<td>55.3 (18.9)</td>
<td>59.2 (18.1)</td>
<td>62.1 (17.4)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>n = 3,839</td>
<td>n = 12,880</td>
<td>n = 5,822</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Age Groups</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;40</td>
<td>935 (24%)</td>
<td>2,070 (16%)</td>
<td>680 (12%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>40-59</td>
<td>1,217 (32%)</td>
<td>4,135 (32%)</td>
<td>1,755 (30%)</td>
<td></td>
</tr>
<tr>
<td>60-69</td>
<td>1,265 (33%)</td>
<td>4,859 (38%)</td>
<td>2,304 (40%)</td>
<td></td>
</tr>
<tr>
<td>80+</td>
<td>422 (11%)</td>
<td>1,816 (14%)</td>
<td>1,083 (19%)</td>
<td></td>
</tr>
<tr>
<td><strong>Gender</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>1,975 (51%)</td>
<td>5,996 (47%)</td>
<td>2,496 (43%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td><strong>Race</strong> Whiteb</td>
<td>3,835 (100%)</td>
<td>12,359 (96%)</td>
<td>5,544 (95%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td><strong>Stage</strong> In Situ</td>
<td>420 (11%)</td>
<td>3,904 (30%)</td>
<td>2,034 (35%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Localized</td>
<td>2,719 (71%)</td>
<td>7,680 (60%)</td>
<td>3,233 (56%)</td>
<td></td>
</tr>
<tr>
<td>Regional</td>
<td>220 (6%)</td>
<td>413 (3%)</td>
<td>262 (5%)</td>
<td></td>
</tr>
<tr>
<td>Distant</td>
<td>220 (6%)</td>
<td>375 (3%)</td>
<td>156 (3%)</td>
<td></td>
</tr>
<tr>
<td>Unstaged</td>
<td>260 (7%)</td>
<td>508 (4%)</td>
<td>137 (2%)</td>
<td></td>
</tr>
<tr>
<td><strong>Morphology In Situ</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superficial Spreading</td>
<td>77 (18%)</td>
<td>631 (16%)</td>
<td>178 (9%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Nodular</td>
<td>1 (0%)c</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td></td>
</tr>
<tr>
<td>Lentigo Maligna</td>
<td>262 (62%)</td>
<td>1,876 (48%)</td>
<td>983 (48%)</td>
<td></td>
</tr>
<tr>
<td>Melanoma NOS</td>
<td>76 (18%)</td>
<td>1,370 (35%)</td>
<td>866 (43%)</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>4 (1%)</td>
<td>27 (1%)</td>
<td>13 (2%)</td>
<td></td>
</tr>
<tr>
<td><strong>Morphology Invasive</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superficial Spreading</td>
<td>1,024 (30%)</td>
<td>4,005 (45%)</td>
<td>1,945 (51%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Nodular</td>
<td>385 (11%)</td>
<td>887 (10%)</td>
<td>394 (10%)</td>
<td></td>
</tr>
<tr>
<td>Lentigo Maligna</td>
<td>185 (5%)</td>
<td>612 (7%)</td>
<td>297 (8%)</td>
<td></td>
</tr>
<tr>
<td>Melanoma NOS</td>
<td>1,684 (49%)</td>
<td>3,135 (35%)</td>
<td>1,000 (26%)</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>141 (4%)</td>
<td>337 (4%)</td>
<td>152 (4%)</td>
<td></td>
</tr>
<tr>
<td><strong>Skin and Sub-sites</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Head/Neck</td>
<td>994 (26%)</td>
<td>3,808 (30%)</td>
<td>1,901 (33%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Trunk</td>
<td>1,061 (28%)</td>
<td>3,514 (27%)</td>
<td>1,488 (26%)</td>
<td></td>
</tr>
<tr>
<td>Upper Limbs</td>
<td>785 (20%)</td>
<td>2,970 (23%)</td>
<td>1,422 (24%)</td>
<td></td>
</tr>
<tr>
<td>Lower Limbs</td>
<td>763 (20%)</td>
<td>2,133 (17%)</td>
<td>843 (14%)</td>
<td></td>
</tr>
<tr>
<td>Skin NOS</td>
<td>236 (6%)</td>
<td>455 (4%)</td>
<td>168 (3%)</td>
<td></td>
</tr>
<tr>
<td><strong>Rurality</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Urban</td>
<td>1,704 (44%)</td>
<td>6,241 (48%)</td>
<td>2,808 (48%)</td>
<td>.0003</td>
</tr>
<tr>
<td>Large Rural</td>
<td>645 (17%)</td>
<td>1,904 (15%)</td>
<td>861 (15%)</td>
<td></td>
</tr>
<tr>
<td>Small Rural</td>
<td>743 (19%)</td>
<td>2,439 (19%)</td>
<td>1,128 (19%)</td>
<td></td>
</tr>
<tr>
<td>Isolated</td>
<td>747 (19%)</td>
<td>2,296 (18%)</td>
<td>1,025 (18%)</td>
<td></td>
</tr>
</tbody>
</table>

*Derived from ANOVA (mean age comparison) and chi-square tests (all other comparisons).

bAll others grouped into non-white or unknown race categories.

cClassification and/or recording error.

significantly from 51% from 1973–1987 down to 43% from 2007–2011. Among the invasive CM cases, a larger proportion were diagnosed with localized disease in the more recent time periods. Throughout the 39-year study period, almost all cases were white, and rurality changed little with just under half of cases residing in urban areas, reflective of Iowa demographics (Table 1).

During 1973–1987, < 3% of invasive CM cases (n = 93) were attributed to private pathology laboratories, increasing to 17% (n = 627) from 2007–2011. Less than 1% (n = 32) of invasive CM cases were attributed to dermatology/private physician offices from 1973–1987, which grew to 3% (n = 124) in the 2007 to 2011 period (Figure 2).

From 1973–1987, 79% (n = 330) of in situ cases were attributed to hospital facilities, 17% (n = 73) to private pathology laboratories, and 4% (n = 17) to dermatology/private physician offices. In the 2007–2011 period, only 55% (n = 1,126) of cases were attributed to hospital facilities, 37% (n = 752) to private pathology laboratories, and 8% (n = 156) to dermatology/private physician offices (Figure 2). For a short time in the period of 1973–1987, the SHRI sent registry staff into a single dermatology office that accounted for all of the in situ cases attributed to dermatology offices during that time period.
In situ cases were attributed to dermatology offices. A higher percentage of urban residents were attributed to nonhospital-based sources compared to hospital-based facilities, likely reflective of urban physicians having more private laboratory options compared to those practicing in rural areas who may rely more on hospital-based pathology laboratories. It should also be noted that Iowa has 82 formally designated critical access hospitals, the second highest in the nation behind Kansas. Critical access–designated hospitals must be located in rural areas over 35 miles from another hospital, or 15 miles from another hospital in mountainous terrain or areas with only secondary roads. Critical access–designated hospitals receive cost-based reimbursement from Medicare in order to improve the financial feasibility of rural hospitals and access to healthcare for rural Americans by reducing rural hospital closures.

**Discussion**

Results indicate that nonhospital-based casefinding sources have been an important contributor to the increasing CM incidence in Iowa, as the incidence rate of invasive CM would have been 20% lower and the in situ rate 44% lower without including them. Private pathology laboratories made the greatest contribution to casefinding, particularly among patients diagnosed with in situ CM who were residing in urban areas. Dermatology offices continue to be a growing source of casefinding, particularly among those diagnosed with in situ CM. Given that rurality is associated with larger proportions of cases reported by hospital-based facilities, likely reflective of urban physicians having more private laboratory options compared to those practicing in rural areas who may rely more on hospital-based pathology laboratories, it is probable that underreporting in more urban populations could be much higher, as has been demonstrated in previous studies.7,10

It should also be noted that Iowa has 82 formally designated critical access hospitals, the second highest in the nation behind Kansas. Critical access–designated hospitals must be located in rural areas over 35 miles from another hospital, or 15 miles from another hospital in mountainous terrain or areas with only secondary roads. Critical access–designated hospitals receive cost-based reimbursement from Medicare in order to improve the financial feasibility of rural hospitals and access to healthcare for rural Americans by reducing rural hospital closures.19 Therefore,

![Figure 2: Percent of Invasive and In Situ Cutaneous Melanoma in Iowa by Reporting Source and Time Period](image)

<table>
<thead>
<tr>
<th>Year</th>
<th>Reporting Source</th>
<th>Invasive</th>
<th>In Situ</th>
</tr>
</thead>
<tbody>
<tr>
<td>1973–1987</td>
<td>Hospitals/Facilities</td>
<td>3,413</td>
<td>420</td>
</tr>
<tr>
<td>1988–2004</td>
<td>Private Path Labs</td>
<td>8,976</td>
<td>420</td>
</tr>
<tr>
<td>2007–2011</td>
<td>Dermatology Offices</td>
<td>7,884</td>
<td>357</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Source</th>
<th>Invasive</th>
<th>In Situ</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hospitals/Facilities</td>
<td>1,817</td>
<td>393</td>
</tr>
<tr>
<td>Private Path Labs</td>
<td>562</td>
<td>299</td>
</tr>
<tr>
<td>Dermatology Offices</td>
<td>741</td>
<td>387</td>
</tr>
</tbody>
</table>

**Figure 3: Percent of Invasive and In Situ Cutaneous Melanoma in Iowa by Reporting Source and Rurality, 2007–2011**

<table>
<thead>
<tr>
<th>Rurality</th>
<th>Reporting Source</th>
<th>Invasive</th>
<th>In Situ</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urban</td>
<td>Hospitals/Facilities</td>
<td>1,817</td>
<td>393</td>
</tr>
<tr>
<td></td>
<td>Private Path Labs</td>
<td>562</td>
<td>299</td>
</tr>
<tr>
<td></td>
<td>Dermatology Offices</td>
<td>741</td>
<td>387</td>
</tr>
<tr>
<td>Large Rural</td>
<td>Hospitals/Facilities</td>
<td>1,258</td>
<td>302</td>
</tr>
<tr>
<td></td>
<td>Private Path Labs</td>
<td>532</td>
<td>271</td>
</tr>
<tr>
<td></td>
<td>Dermatology Offices</td>
<td>560</td>
<td>325</td>
</tr>
<tr>
<td>Small Rural</td>
<td>Hospitals/Facilities</td>
<td>1,086</td>
<td>267</td>
</tr>
<tr>
<td></td>
<td>Private Path Labs</td>
<td>506</td>
<td>253</td>
</tr>
<tr>
<td></td>
<td>Dermatology Offices</td>
<td>524</td>
<td>297</td>
</tr>
<tr>
<td>Isolated</td>
<td>Hospitals/Facilities</td>
<td>1,554</td>
<td>409</td>
</tr>
<tr>
<td></td>
<td>Private Path Labs</td>
<td>520</td>
<td>289</td>
</tr>
<tr>
<td></td>
<td>Dermatology Offices</td>
<td>531</td>
<td>320</td>
</tr>
</tbody>
</table>

Approximately two-thirds of the 68 active dermatologists in Iowa. Based on these results and the following assumptions: 1) constant volume of CM diagnoses across all dermatologists practicing in Iowa, 2) constant proportion of cases that would not be detected through hospitals and private pathology laboratories, and 3) the two-thirds of dermatologists currently reporting to the SHRI reported all of their CM cases during this time period, we estimate that the one-third not reporting may have diagnosed/treated approximately 124 CM cases that were never reported to the SHRI.
the relatively large number of rural hospitals in Iowa also likely further explains the larger proportion of rural cases reported from hospital as opposed to nonhospital sources.

A majority of missed cases are most likely in situ and localized CM. In situ CM was a fairly small proportion of all cases during 1973–1987, but made up over a third of all cases by the most recent time period. Increased surveillance, diagnosis, and reporting of lentigo maligna, a common subtype of in situ CM that occurs on chronically sun-exposed locations in elderly patients, may explain the increasing age at diagnosis across the 3 time periods, as well as the increasing proportion of lesions occurring on the head/neck.20 While we found higher proportions of lentigo maligna than have been previously reported in earlier time periods (1980s), incidence has likely been underestimated due to underreporting, and the proportion of lentigo maligna would be expected to increase as the population ages and undergoes increased clinical surveillance.21

While efforts to reach out to dermatologists did not yield a high volume of new cases relative to traditional surveillance methods, it is possible that there were more cases never captured by SHRI that could have been attributed to dermatology offices if all were routinely reporting their CM cases. As new independent laboratories continue to enter the marketplace and an increasing proportion of patients are treated in the outpatient setting, direct reporting to registries by dermatology offices through the Center for Medicare and Medicaid Services Meaningful Use initiatives may provide valuable casefinding information to registries. This reporting could include pathology

Table 2. Characteristics of Cases of Cutaneous Melanoma Ascertained in Iowa in 2007–2011 by Reporting Source

<table>
<thead>
<tr>
<th>Reporting Source</th>
<th>Hospital-Based Reporting Sources</th>
<th>Private Path Labs and Dermatology Offices</th>
<th>P*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Mean (SD)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;40</td>
<td>497 (12%)</td>
<td>182 (11%)</td>
<td>.3331</td>
</tr>
<tr>
<td>40-59</td>
<td>1,246 (30%)</td>
<td>508 (31%)</td>
<td></td>
</tr>
<tr>
<td>60-69</td>
<td>1,661 (40%)</td>
<td>642 (39%)</td>
<td></td>
</tr>
<tr>
<td>80+</td>
<td>750 (18%)</td>
<td>327 (20%)</td>
<td></td>
</tr>
<tr>
<td>Gender</td>
<td>Female</td>
<td>1,742 (42%)</td>
<td>.0726</td>
</tr>
<tr>
<td>Race</td>
<td>White</td>
<td>4,102 (99%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Stage</td>
<td>In Situ</td>
<td>1,126 (27%)</td>
<td></td>
</tr>
<tr>
<td>Localized</td>
<td>2,557 (62%)</td>
<td>676 (41%)</td>
<td></td>
</tr>
<tr>
<td>Regional</td>
<td>260 (6%)</td>
<td>2 (0%)</td>
<td></td>
</tr>
<tr>
<td>Distant</td>
<td>156 (4%)</td>
<td>0 (0%)</td>
<td></td>
</tr>
<tr>
<td>Unstaged</td>
<td>55 (1%)</td>
<td>73 (4%)</td>
<td></td>
</tr>
<tr>
<td>Morphology In Situ</td>
<td>Superficial Spreading</td>
<td>96 (9%)</td>
<td>.055</td>
</tr>
<tr>
<td>Lentigo Maligna</td>
<td>509 (45%)</td>
<td>474 (52%)</td>
<td></td>
</tr>
<tr>
<td>NOS</td>
<td>518 (46%)</td>
<td>348 (38%)</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>3 (0%)</td>
<td>4 (0%)</td>
<td></td>
</tr>
<tr>
<td>Morphology Invasive</td>
<td>Superficial Spreading</td>
<td>1,502 (50%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Nodular</td>
<td>371 (12%)</td>
<td>23 (3%)</td>
<td></td>
</tr>
<tr>
<td>Lentigo Maligna</td>
<td>222 (7%)</td>
<td>75 (10%)</td>
<td></td>
</tr>
<tr>
<td>NOS</td>
<td>800 (26%)</td>
<td>191 (25%)</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>133 (4%)</td>
<td>19 (3%)</td>
<td></td>
</tr>
<tr>
<td>Skin and Sub-sites</td>
<td>Head/Neck</td>
<td>1,314 (32%)</td>
<td>.0004</td>
</tr>
<tr>
<td>Trunk</td>
<td>1,087 (26%)</td>
<td>400 (24%)</td>
<td></td>
</tr>
<tr>
<td>Upper Limbs</td>
<td>1,000 (24%)</td>
<td>422 (25%)</td>
<td></td>
</tr>
<tr>
<td>Lower Limbs</td>
<td>619 (15%)</td>
<td>224 (14%)</td>
<td></td>
</tr>
<tr>
<td>Skin NOS</td>
<td>134 (3%)</td>
<td>27 (2%)</td>
<td></td>
</tr>
<tr>
<td>Rurality</td>
<td>Urban</td>
<td>1,866 (45%)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Large Rural</td>
<td>623 (15%)</td>
<td>237 (14%)</td>
<td></td>
</tr>
<tr>
<td>Small Rural</td>
<td>848 (20%)</td>
<td>278 (17%)</td>
<td></td>
</tr>
<tr>
<td>Isolated</td>
<td>817 (20%)</td>
<td>206 (12%)</td>
<td></td>
</tr>
</tbody>
</table>

aDerived from T-test (mean age comparison) and chi-square tests (all other comparisons).

bAll others grouped into non-white or unknown race categories.
Figure 4. State Health Registry of Iowa Melanoma Reporting Form

<table>
<thead>
<tr>
<th>Patient First Name: ______________________</th>
<th>Middle: ______________________</th>
<th>Last: ______________________</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date of Birth: ______________________</td>
<td>SS#: <strong><strong><strong>-</strong><strong>-</strong></strong></strong></td>
<td>Sex: Male:____ Female:____</td>
</tr>
<tr>
<td>Race: Caucasian:____ African American:____ Native American:____ Other: ______________________</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ethnicity: Hispanic/Spanish? Yes:____ No:____ Unknown:____</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Insurance Provider: ______________________</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patient Address: ______________________</td>
<td>Street: __________________________________________________________________</td>
<td></td>
</tr>
<tr>
<td></td>
<td>City: ________________________</td>
<td>State: __________ Zip: ______</td>
</tr>
<tr>
<td>Marital Status: Single:____ Married:_____ Divorced:____ Divorced:____ Widowed:____ Unknown:____</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Date of Diagnosis:____________ Dermatologist who diagnosed the melanoma: ______________________</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Site: ______________________ Laterality: Right:____ Left:____ Midline:____ Unknown:____</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulceration Present: Yes:____ No:____ Maximum tumor size:________________</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

| Was there lymph node involvement? | Yes:____ No:____ Unknown:____ |
| Satellite nodule? | Yes:____ No:____ Unknown:____ |
| In-transit metastasis? | Yes:____ No:____ Unknown:____ |
| Sentinel node bx performed? | Yes:____ No:____ Unknown:____ |

| Serum LDH test done: | Yes:____ Results within normal limits:____ Results elevated:____ |
| No Serum LDH test done:____ Unknown:____ if Serum LDH test done:____ |

Treatment received for this melanoma:

| Surgery: | Biopsy:____ Excision:____ Wide/re-excision:____ Dates:________________________________________ |
| Radiation: | No:____ Yes - Facility:____________________ Date:________________ |
| Chemotherapy: | No:____ Yes - Type:____________ Facility:________________ Date:_________ |
| Immunotherapy: | No:____ Yes - Type:____________ Facility:________________ Date:_________ |

Does this patient have a history of previous melanoma?

| Yes, but current melanoma is a new primary:____ | Yes, current melanoma is a recurrence:____ |
| No history of previous melanoma:____ Unknown if history of previous melanoma:____ |

Dermatologist completing form: ______________________ Phone Number: (______)______-______

Please FAX the completed form and all pertinent path reports to the State Health Registry of Iowa Attention: Rod Burnett. Fax #: 319-335-8610.

If you have any questions please call Rod Burnett at 319-384-3226.

Thank you for your time and effort to assure complete and accurate reporting of Melanoma Form 1212.
report information from independent laboratories not known to the registries (otherwise missed). Furthermore, cancer reporting by dermatologists can provide important treatment information necessary for CM population-based studies otherwise inaccessible to registries. As previous studies have shown that many dermatologists are unaware they should be reporting CM cases to their state’s central cancer registry,\textsuperscript{7,8} electronic reporting of CM to fulfill Meaningful Use measures could be an important step toward more complete case ascertainment.

A complementary effort to incorporate an educational practice improvement component related to CM reporting requirements, such as those suggested by The American Board of Dermatology Maintenance of Certification Program Component 4, which requires practice performance evaluation including completion of a practice assessment/quality improvement program and peer and patient surveys, may also be effective in promoting case reporting by dermatology offices.\textsuperscript{9} This type of educational component could include specific information on how relatively simple case report forms (Figure 4) can be completed and sent to the state’s central cancer registry (along with pathology reports), where the registry’s abstractors can use the submitted information to prepare a formal abstract. Either approach would likely enhance casefinding beyond current labor-intensive efforts of registries to track down cases from nonhospital reporting sources.

There are several limitations to consider when interpreting the results of this paper. First, we had to collapse a number of reporting sources into the hospital facility category (eg, ambulatory surgery centers) because some were not separately categorized throughout the study period, and some yielded too few cases to evaluate separately. Also, a single state registry was examined and may not be generalizable to all geographic locations, although it is likely that evolving reporting sources outside of hospital facilities are an issue for all population-based registries. Furthermore, the pattern of increasing incidence of CM is occurring among almost all SEER registries.\textsuperscript{10} Unfortunately, as the SEER database does not contain CM risk factor information such as personal lifetime UV exposure, we were unable to determine what proportion of the increase in CM over the time period is due to more complete reporting as opposed to increased UV exposure and/or other risk factors, or alternatively, increased awareness among both health care providers and the general public leading to more CM diagnoses. Finally, as nonhospital based reporting sources are more likely to report race as “unknown,” it is possible the proportion of whites attributed to nonhospital based reporting sources is underreported. This limitation will become increasingly important to future registry-based research as central registries receive more case reports from nonhospital sources lacking race information.

In conclusion, accurate and thorough reporting of CM cases is important to researchers examining ways to prevent and treat CM, as well as to policy makers who need to target programs and funding to populations most in need. Initiatives designed to encourage and facilitate reporting from private pathology laboratories and dermatology offices will likely yield increased melanoma casefinding in the United States, particularly for in situ CM cases. However, aggressively pursuing cases from dermatology offices can be a resource intensive endeavor for cancer registries. It is possible that the activities and incentives that promote practice improvement and electronic reporting of cancer cases and associated treatment information to state cancer registries may in the future lead to increased efficiency of reporting of cases diagnosed and treated in physician offices. Further evaluation of the impact of new reporting sources on the incidence of CM will continue to be important, especially with the adoption of electronic health record implementation and the Meaningful Use initiative related to electronic reporting to cancer registries.

References


The Facility Oncology Registry Data Standards, better known as FORDS, were developed in 2003 by the Commission on Cancer (CoC) of the American College of Surgeons for its CoC-accredited programs. Although updated periodically to ensure that appropriate codes were being used by registrars in CoC-accredited facilities, there has not been any major overhaul of the manual since inception. The FORDS manual replaces ROADS (Registry Operations and Data Standards) which was originally implemented in 1996. The FORDS rules for coding are important not only to ensure that appropriate data are placed into our hospital registry abstracts, but also to support the important activities of the National Cancer Data Base (NCDB) first implemented in the late 1980s by the American College of Surgeons and the American Cancer Society. The NCDB, which now includes over 30 million accessions, is a vital tool not only for comparison of data, but also for development of new strategies for cancer staging.

Material that is placed into cancer registry abstracts as defined by FORDS also determines the CoC quality measures currently being used in the Rapid Quality Reporting System, Cancer Quality Improvement Program, and the CP3R Program, which tracks quality within all of the CoC-accredited institutions. As mentioned, the NCDB is a powerful research tool. The Participant User File is now used by many as a rich clinical research tool to enhance cancer care.

Changes in the FORDS manual also affect other standards and imply important changes for other members of the cancer surveillance community. The National Program of Cancer Registries under the auspices of the Centers for Disease Control and Prevention (CDC) and the Surveillance, Epidemiology, and End Results (SEER) program of the National Cancer Institute are affected by revisions in FORDS and, therefore, individuals who are leaders in these programs are included in the FORDS revision leadership. In addition, the eventual revision with changes to the FORDS process must be sanctioned by the North American Association of Central Cancer Registries (NAACCR).

To enhance the revision of the FORDS manual, a multidisciplinary Steering Committee has been developed to lead this overall process and to assure that all voices are heard during the revision of codes that affect many segments of cancer care in United States. These members of the Steering Committee will ensure that in the years to come, complete clinical and pathological indicators are collected for proper cancer staging. We need to collect better recurrence data; this will be achieved by adding appropriate codes to the document. New systemic cancer treatments will also be captured in appropriate codes added to the FORDS process. Finally, codes that are no longer appropriate must be eliminated to make registrar workload more efficient by avoiding the continuation of extraneous and inappropriate codes that are no longer relevant in cancer treatment.

The FORDS revision will not occur in a vacuum. Many other activities will be ongoing over the next 2 to 3 years that have significant implications regarding the FORDS revision process. The Collaborative Stage program currently sponsored by the CDC and administered through the American Joint Committee on Cancer (AJCC) will end in 2015 and, therefore, all standard setters are being trained into the collection of AJCC clinical and pathologic TNM classifications and stage group data during this time frame. In addition, electronic health record changes and harmonization of data collection under NAACCR will be implemented and the likely introduction of ICD-10 will obviously be another factor in this process.

To ensure appropriate and effective revision of all codes, clinicians will be heavily involved in the process and will be invited to participate through membership in expert panels that have been created for the development of the eighth edition of the AJCC Cancer Staging Manual that will be completed by the fall of 2016. An instructional webinar has been developed for clinicians involved in the FORDS revision process, and it is anticipated that all cancer sites will be reviewed for correctness of coding and for the inclusion of data fields and codes that reflect modern cancer care. To ensure that clinicians, registrars and all interested parties have a voice in this process, a website has been developed to solicit recommendations for addition, deletion, or change of existing codes.

The FORDS revision process may be the most important project that has occurred in the surveillance and collection of data relating to treatment of cancer patients in the United States. This process will assure that data used for modern cancer treatment is collected appropriately and utilized by all members of the treatment and surveillance community. It goes without saying that the National Cancer Registrars Association and its members are an integral and vital part of this process.
PROCESS IMPROVEMENT: A MULTI-REGISTRY DATABASE ABSTRACTION SUCCESS STORY

Quiz Instructions: The multiple choice or true/false quiz below is provided as an alternative method of earning CE credit hours. Refer to the article for the ONE best answer to each question. The questions are based solely on the content of the article. Answer the questions and send the original quiz answer sheet and fee to the NCRA Executive Office before the processing date listed on the answer sheet. Quizzes may not be retaken nor can NCRA staff respond to questions regarding answers. Allow 4–6 weeks for processing following the submission deadline to receive return notification of your completion of the CE process. The CE hour will be dated when it is submitted for grading; that date will determine the CE cycle year.

After reading this article and taking the quiz, the participants will be able to:
• Describe problems related to delayed chart abstraction
• Identify methods to improve productivity
• Discuss how timely abstracting can improve patient care

1. National reporting of core measures with respect to cardiac care enables participating institutions to:

<table>
<thead>
<tr>
<th>gauge performance compared to national averages</th>
<th>justify staffing increases in the registry</th>
<th>derive financial benefits by adhering to core measures</th>
<th>utilize benchmarks for quality improvement efforts</th>
</tr>
</thead>
<tbody>
<tr>
<td>a) Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
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<td>b) Y</td>
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<td>c) Y</td>
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<td>Y</td>
</tr>
<tr>
<td>d) N</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
</tr>
</tbody>
</table>

2. Delayed retrospective chart abstraction results in the:
   a) timely prescription of appropriate medications by providers
   b) timely identification of missed opportunities of care
   c) ability to amend the medical record according to hospital regulations
   d) potential of financial loss due to late adherence to core measures

3. This process improvement initiative was created in order to:
   a) justify overtime for abstractors
   b) validate the need for additional full-time equivalents (FTEs)
   c) reduce backlog while keeping up with daily demands
   d) provide bonuses to providers who consistently meet core measures

4. In which step of the DMAIC (define, measure, analyze, improve, control) process improvement plan are methods to improve abstraction efficiency and reduce backlog identified?
   a) analyze
   b) control
   c) define
   d) improve

5. The employees working in the Cardiac Database Registry:
   a) lacked experience in health information data abstraction
   b) were cross-trained with equal division of abstraction
   c) needed module-specific knowledge
   d) were paid on a per case completed basis

6. Daily capacity was calculated for 4 employees, assuming which of the following?
   a) 8 hours per day dedicated to abstracting
   b) 6 hours per day dedicated to abstracting
   c) 1 hour per day for lunch
   d) 0.5 hour per day for other work

7. According to Table 2, Daily Abstraction Demand:
   a) STS National Database abstracts require the greatest number of minutes per case
   b) CathPCI Registry (percutaneous coronary interventions) abstracts require the greatest number of minutes per case
   c) STS National Database abstracts require the greatest total hours for all cases
   d) CathPCI Registry (percutaneous coronary interventions) abstracts require the greatest total hours for all cases

8. According to Table 5, Required Number of Cases to be Abstracted Daily to Reach Goal, abstractors must complete ___ backlogged cases daily to reach the goal.
   a) 6
   b) 8
   c) 16
   d) 24

9. Challenges associated with this process improvement initiative include:
   a) higher employee productivity
   b) employees’ resistance to change
   c) introduction of work accountability
   d) new expectations for employee work output

<table>
<thead>
<tr>
<th>higher employee productivity</th>
<th>employees’ resistance to change</th>
<th>introduction of work accountability</th>
<th>new expectations for employee work output</th>
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<td>b) Y</td>
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<td>c) Y</td>
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<td>Y</td>
</tr>
<tr>
<td>d) N</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
</tr>
</tbody>
</table>

10. This registry was able to reduce backlog abstracting by:
   a) using measurement-driven work output
   b) cancelling vacation time
   c) authorizing overtime
   d) hiring additional FTEs

The JRM Quiz and answers are now available through NCRA’s Center for Cancer Registry Education (CCRE). For your convenience, the JRM article and quiz can be accessed online at www.CancerRegistryEducation.org/jrm-quizzes. Download the article, complete the quiz and claim CE credit all online.
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1   A  B  C  D
2   A  B  C  D
3   A  B  C  D
4   A  B  C  D
5   A  B  C  D
6   A  B  C  D
7   A  B  C  D
8   A  B  C  D
9   A  B  C  D
10  A  B  C  D

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No photocopies will be accepted.

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March 31, 2016

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What Increased Registry Outreach may mean for a Cutaneous Melanoma Surveillance: Impact of Changes in Iowa.


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Vicki G. Nelson, MPH, RHIT, CTR | EDITOR-IN-CHIEF, JRM

The Journal of Registry Management, official journal of the National Cancer Registrars Association (NCRA), announces a call for original manuscripts on registry methodology or research findings related to the 5 subjects listed below and related topics.

Topics:
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Contributed manuscripts are peer-reviewed prior to publication.
Manuscripts of the following types may be submitted for publication:
1. Methodology Articles addressing topics of broad interest and appeal to the readership, including methodological aspects of registry organization and operation.
2. Research articles reporting findings of original, reviewed, data-based research.
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5. Opinion papers/editorials including position papers, commentaries, essays, and interviews that analyze current or controversial issues and provide creative, reflective treatments of topics related to registry management.
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Address all manuscripts to: Vicki G. Nelson, MPH, RHIT, CTR, Editor-in-Chief, Journal of Registry Management, (770) 488-6490, vnelson@cdc.gov.

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Manuscripts may be submitted for publication in the following categories: Articles addressing topics of broad interest and appeal to the readership, including Methodology papers about registry organization and operation, Research papers reporting findings of original, reviewed, data-based research; Primers providing tutorials on relevant subjects; and “How I Do It” papers are also solicited. Opinion papers/editorials including position papers, commentaries, and essays that analyze current or controversial issues and provide creative, reflective treatments of topics related to registry management; Letters to the Editor, and specifically-targeted Bibliographies of significant interest are invited.

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